RADIOLOGY OF LIVER CIRCULATION

SERIES IN RADIOLOGY

J. Odo Op den Orth, The Standard Biphasic-Contrast Examination of the Stomach and Duodenum: Method, Results and Radiological Atlas.
1979. ISBN 90 247 2159 8

J.L. Sellink and R.E. Miller, Radiology of the Small Bowel. Modern Enteroclysis Technique and Atlas.
1981. ISBN 90 247 2460 0

R.E. Miller and J. Skucas, The Radiological Examination of the Colon. Practical Diagnosis.
1983. ISBN 90 247 2666 2

S. Forgács, Bones and Joints in Diabetes Melitus.
1982. ISBN 90 247 2395 7

G. Németh and H. Kuttig, Isodose Atlas. For Use in Radiotherapy.
1981. ISBN 90 247 2476 7

J. Chermet, Atlas of Phlebography of the Lower Limbs, including the Iliac Veins.
1982. ISBN 90 247 2525 9

B. Janevski, Angiography of the Upper Extremity.
1982. ISBN 90 247 2684 0

M.A.M. Feldberg, Computed Tomography of the Retroperitoneum. An Anatomical and Pathological Atlas with Emphasis on the Fascial Planes.
1983. ISBN 0 89838 573 3

L.E.H. Lampmann, S.A. Duursma and J.H.J. Ruys, CT Densitometry in Osteoporosis. The Impact on Management of the Patient.
1984. ISBN 0 89838 633 0

J.J. Broerse and T.J. MacVittie, Response of Different Species to Total Body Irradiation.
1984. ISBN 0 89838 678 0

C. L'Herminé, Radiology of Liver Circulation.
1985. ISBN 0 89838 715 9

RADIOLOGY OF LIVER CIRCULATION

CLAUDE L'HERMINÉ M.D.
Professor of Radiology
University of Lille Medical School
Lille, France

1985 **MARTINUS NIJHOFF PUBLISHERS**
a member of the KLUWER ACADEMIC PUBLISHERS GROUP
BOSTON / DORDRECHT / LANCASTER

Distributors

for the United States and Canada: Kluwer Academic Publishers, 190 Old Derby Street, Hingham, MA 02043, USA
for the UK and Ireland: Kluwer Academic Publishers, MTP Press Limited, Falcon House, Queen Square, Lancaster LA1 1RN, UK
for all other countries: Kluwer Academic Publishers Group, Distribution Center, P.O. Box 322, 3300 AH Dordrecht, The Netherlands

Library of Congress Cataloging in Publication Data

L'Herminé, Claude.
Radiology of liver circulation.

(Series in radiology)
Bibliography: p.
Includes index.
1. Liver--Blood-vessels--Radiography. 2. Liver--Diseases--Diagnosis. 3. Portal hypertension--Diagnosis. I. Title. II. Series. [DNLM: 1. Liver--radiography. 2. Liver Circulation. WI 702 L688r]
RC847.H38 1985 616.3'620757 85-8905

ISBN-13: 978-94-010-8718-6 e-ISBN-13: 978-94-009-5010-8
DOI: 10.1007/978-94-009-5010-8

Copyright

TABLE OF CONTENTS

INTRODUCTION

The liver blood flow disorders are usually described under the term 'portal hypertension', which is a well-known syndrome since it has been widely studied for years from a clinical, radiological and therapeutic point of view.

In fact, portal hypertension is only the result of an obstacle to the portal flow, the clinical manifestations of which often occur at a late stage of the disease: bleeding esophageal varices, splenomegaly, ascites.

In addition, it is well known that there is not always a precise relationship between the degree of the obstruction, i.e. the decrease of the portal inflow and the level of the portal hypertension. A severe obstruction to the portal flow may therefore sometimes remain clinically inconspicuous for a long time.

For this reason improvement of our knowledge of portal hypertension mainly relies on angiographic investigations. Thus, splenoportography permits the discovery of prehepatic obstructions whereas hepatic venography permitted the recognition of two different types of intrahepatic obstruction according to the pre-sinusoidal or post-sinusoidal site of the obstacle. More recently, arterial hepatic changes could be evidenced through arteriography.

However, these angiographic investigations have not yet allowed for all the problems about portal hypertension to be clearly identified.

There are probably two main reasons for this; first, each angiographic method does not permit the documentation of all the vascular structures of the liver so that only partial information is obtained in most cases. Second, angiographic abnormalities have been appreciated mainly from a morphological point of view whereas too little attention has been paid to the hemodynamical significance of the angiographic signs. However, arterioportography appears to be an accurate and reliable method for evaluating hepatic blood flow disturbances when considering that every evidenced morphological change does have a precise hemodynamical significance.

Arteriography therefore permits the evaluation of the degree of the decreased portal inflow to the liver which is the main abnormality leading to portal hypertension, as well as a demonstration of the site of the obstruction, its progression rate, and sometimes even its cause.

Consequently, many obscure points about portal hypertension may be elicited and responses may be given to some questions which have up now not been clearly responded to, such as: What does splenomegaly mean in portal hypertension? Does portal hypertension without obstruction exist? What is the role of the hepatic artery? In the same way, intrahepatic obstructions to the portal flow without portal hypertension may be demonstred. Information about hemodynamics resulting from arterioportography is all the more useful to the diagnosis, the treatment and a better understanding of portal hypertension as few other reliable methods may be resorted to: The clinical manifestations are nonspecific and often delayed signs; there is no possibility for the portal flow rate to be measured in clinical conditions; from a pathological point of view, the percutaneous or transjugular liver biopsy itself is not always reliable.

For these reasons, it is worth evaluating as precisely as possible the hemodynamics of the liver blood flow on the basis of arterioportography since it may be helpful in anticipating the main features of the pathological process involving the liver parenchyma as well as in making the right diagnosis.

ACKNOWLEDGEMENTS

In order to demonstrate that a better understanding of the hemodynamic significance of the angiographic signs permits a reliable evaluation of liver circulation abnormalities, arterioportography was correlated with clinical and biological data together with pressure recording, liver biopsy, surgical findings and, in some cases, long-term follow-up.

This work was performed in the Department of Radiology of Lille University Hospital on the basis of more than one thousand patients studied over about fifteen years.

Many people have thus contributed to this study in one way or another.

First of all, I would like to thank my friend Prof. J.C. Paris, Chief of the Department of Gastroenterology whose cooperation and interest in this problem allowed us to attain our goal.

I am also indebted to my colleagues Prof. Dr. M. Ribet, Prof. Dr. P. Quandalle and Prof. Dr. M. Vankemmel, surgeons at the Lille University Hospital for their advice and cooperation.

A special acknowledgement is further due to the radiologists who helped me in this research, in particular Prof. Dr. G. Lemaitre, Chairman of the Department of Radiology, and Dr. J.L. Dehaene, Dr. O. Delemazure and Dr. P. Chastanet.

To those who contributed to the writing and production of this book, that is, Mrs. R.M. Dumoulin, for the typing of the manuscript, to Mr. Berteloot for the translation, to Mr. Delattre for the photographs, and, to my publishers for their careful attention to this volume I would like to express my gratitude.

Finally I would like to thank all of those who would agree to combining our results with their own, for only then will further improvement in our knowledge of liver circulation abnormalities be possible.

1. NORMAL HEPATIC CIRCULATION – ANATOMY AND PHYSIOLOGY

I. The hepatic lobule and its three vascular axes

On histological sections, the hepatic lobule has been described as the anatomical unit of the liver. It is made up of cords of liver cells surrounded by the blood sinusoids radiating from the periphery of the lobule towards a central vein. The sinusoids are supplied with blood by portal venules and hepatic artery branches running in the interlobular spaces or portal tracts at the periphery of the lobules.

The blood flows through the sinusoids from the periphery towards the centre where it drains into the central vein. The central veins unit to sublobular veins which, in turn form the roots of the hepatic veins and finally join the inferior vena cava. The hepatic artery branches give rise to perilobular arteries running with the portal venules and biliary ductules in the portal triad and draining into the peripheral part of the sinusoids. This anatomical structure of the hepatic lobule may be documented by microangiographic studies, as demonstrated for instance by Chenderovitch (1).

However, when the structure of the liver is considered in a three-dimensional way, it appears, according to Elias (2, 3) as a sponge-like wallwork of cellular mass tunneled by a communicating system of cavities which contain the blood capillary of the liver. Thus, the sinusoid runs between irregular walls made up of a network of one cell-thick liver cell plates.

Therefore, the functional unit of the liver is described nowadays as the acinus: a mass of hepatocytes distributed around an axis consisting of a terminal hepatic arteriole, portal veinule, bile duct and bordered with several terminal hepatic venules (central veins) which are supplied with blood from several acini

II. Intrahepatic arterio-portal communications

While hepatic artery-portal vein shunting within the cirrhotic liver is well demonstrated on the basis of pathologic and angiographic studies, there is controversy as to whether such communications exist in the normal liver. According to certain authors, presinusoidal hepatic artery-portal vein shunting does not exist in the normal liver, the terminal arterioles ending into intralobular or perilobular sinusoids, and what is more the peribiliary arterial plexus does not apparently communicate with portal veins (3). However, some anatomical studies in humans and experimental works in animals have demonstrated such presinusoidal hepatic artery-portal vein shunting to be present in the normal liver (4, 5, 6, 7, 8).

By microangiography Chenderovitch observed that injection of contrast medium into the hepatic artery results in sinusoidal filling in the same way as injection in the portal vein. On some histological sections hepatic arterioles are seen to end into the sinusoids at the periphery of the hepatic lobule. But on some others the perilobular arteriole empties directly into the perilobular portal veins in a termino-lateral way. This is probably the reason why a retrograde filling of portal veins is frequently seen after injection of contrast medium in the hepatic artery on the microangiographic sections (1). In addition, hepatic artery-portal vein shunting is frequently demonstrated in the subcapsular region of the liver where there exists a very dense arterial and portal venous network. Finally, Chenderovitch thinks that peribiliary arterial plexus drains into the portal veins.

In the same way, Platteborse in 1971 and Osteaux and Jeanmart in 1976 elicited these arterio-portal communications by studying in vivo the hepatic microcirculation in normal animals by injecting either colorating substances or contrast medium into the blood stream (9, 10). In both cases a hemodynamical study could be achieved, demonstrating that the arterial blood reached the sinusoids either directly (arterio-sinusoidal flow) or indirectly through portal venules (arterio-porto-sinusoidal

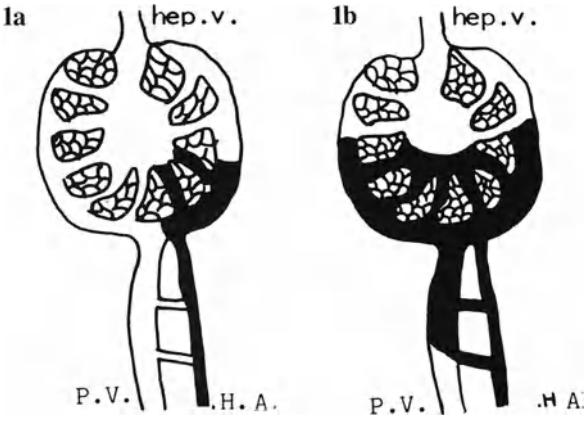


Fig. 1. a/b – Schematic drawing of the hepatic microcirculation after injecting contrast medium in the hepatic artery according to Osteaux and Jeanmart (9) (with permission of Journal de Radiologie)

Ia: Arterio-sinusoidal phase.

Ib: Arterio-porto-sinusoidal phase.

(P.V. = portal vein, H.A. = hepatic artery, hep.v. = hepatic vein)

flow). In some cases the arterioporto-sinusoidal flow appeared to be predominant as compared to arterio-sinusoidal flow (Fig. 1) (9).

These works seem to demonstrate that there exist arterio-portal anastomoses in the normal liver, which were also elicited through anatomical and experimental studies (7, 8, 11, 12). From a physiological point of view, these arterio-portal communications probably have little importance, the arterial and portal venous blood flowing together through the sinusoids towards the hepatic veins. However, in case a markedly decreased portal inflow is associated with an increased arterial flow, such anastomoses could give rise to massive arterio-portal shunting. This phenomenon has been demonstrated in the dog by Lunderquist: after obstruction of the portal vein with a balloon catheter, selective hepatic arteriography resulted in retrograde opacification of the portal vein (13).

In conclusion, whatever the precise site and structure of these anatomical pathways allowing the hepatic arterial blood to reach the small intrahepatic portal veins, these experimental works suggest that hepatic artery may play an important role in the blood supply to normal hepatic parenchyma and account for the relative frequency of arterio-portal shunting as demonstrated by arteriography, not only in cirrhosis but in other circumstances such as liver trauma (Chapter VIII).

III. Normal hepatic circulation

Hepatic circulation is a three-way system: two inflow ways, the portal vein and hepatic artery and one outflow tract, the hepatic veins (Fig. 2). In the normal person, 70 to 80 per cent of the total hepatic blood flow comes from the portal vein; the remainder comes from the hepatic artery.

Another important feature of the hepatic circulation is that a high portal flow rate is associated with a low pressure gradient. The normal portal pressure which is the same as the wedged hepatic-vein pressure ranges from 4 to 11 mm Hg, while the free hepatic vein pressure is the same as the inferior vena cava pressure. The gradient between wedged hepatic vein pressure and inferior vena cava pressure is called the corrected sinusoidal pressure (C.S.P.). It ranges normally from 4 to 7 mm Hg.

Despite this low pressure gradient, the portal flow rate is as high as 1200 ml/min, which means that the intrahepatic vascular resistances are normally very low. As a result, it can be inferred that minimal hepatic lesions may be able to increase the vascular resistances, giving rise to moderate and clinically untraceable portal hypertension, provided the intrahepatic venous bed is diffusely involved.

There is little occurrence of portal hypertension associated with minimal intra-hepatic lesions except in a few cases of liver steatosis or incipiens virus hepatitis for instance. However, it is probably a frequent but unrecognized condition because either no clinical sign of portal hypertension is experienced by the patient or percutaneous liver biopsy is not always reliable enough. Indeed, when a patient is referred to us with portal hypertension we never know exactly when the disease began, and it is likely that in many slowly progessive cases the first stages of the disease remain for a long time clinically unknown.

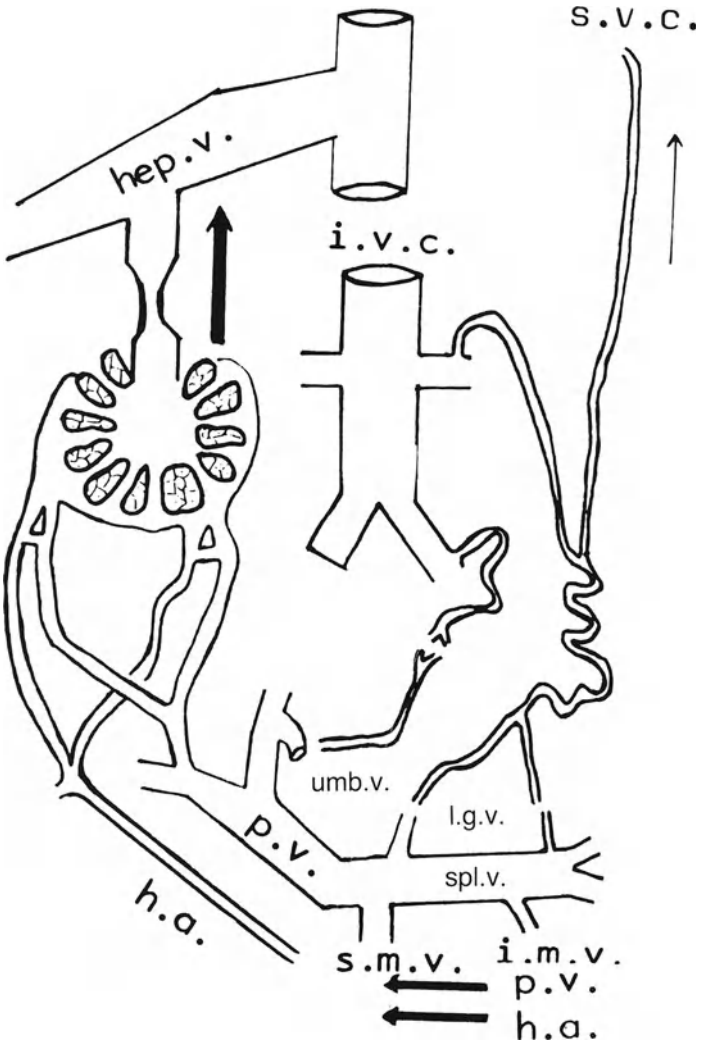


Fig. 2. Schematic drawing of the liver circulation. Under normal conditions the hepatic lobule has two afferent ways, the portal vein and the hepatic artery and one efferent way. In case of post-sinusoidal obstruction it becomes a four-way circulatory system: two afferent ways, portal vein and hepatic artery and two efferent ways, hepatic veins and hepatofugal porto-systemic collaterals. (h.a. = hepatic artery, p.v. = portal vein, i.v.m. = inferior mesenteric vein, s.m.v. = superior mesenteric vein, spl.v. = splenic vein, l.g.v. = left gastric vein, umb.v. = umbilical vein, i.v.c. = inferior vena cava, s.v.c. = superior vena cava)

2. ANGIOGRAPHIC METHODS

I. Direct portal venous opacification

For many years, splenoportography was the primary and only angiographic method used to visualize the portal vein. But other methods such as umbilical portography, percutaneous transhepatic portography, and peroperative ileoportography are also available.

Direct access to the portal venous bed allows for the measuring of portal pressure and for better opacification of the collateral circulation. These are the two main advantages of these methods (14). However, the level of the portal pressure is not always a reliable criterion to evaluate the severity of the disease. It does not closely correlate with the increase of intrahepatic vascular resistances but depends on the development of the collateral circulation as well.

In advanced cirrhosis, for instance, a severe obstruction to the portal inflow may be associated with only moderate increase of the portal pressure, provided a high hepatofugal flow rate has developed through large porto-systemic collateral veins. The collateral circulation is but a consequence of portal hypertension and a precise demonstration of the whole portal-systemic collateral net-work has little practical interest in most cases.

An evaluation of the hepatofugal flow rate through these collateral veins would be more interesting but does not seem to be reliable on the basis of non-physiological methods. Another drawback is the fact that only one of the three vascular axes of the liver can be documented by these methods. They have therefore only little interest from a hemodynamical point of view.

II. Hepatic venous study

It enables the sinusoidal pressure to be recorded through a catheter wedged in a peripheral hepatic vein, as well as the free hepatic vein pressure. Wedged hepatic venous pressure minus free hepatic venous pressure is the corrected sinusoidal pressure which correlates with intrahepatic vascular resistances.

Hepatic vein opacification provides information about the morphological changes of these vessels but its value is questionable from a hemodynamical point of view. The results of wedged hepatic venography depent greatly on the precise position of the catheter tip, the volume of contrast medium, and the injection rate (15, 16, 17). For instance, too rapid an injection results in reflux of contrast medium into portal veins in normal persons, whereas this phenomenon is described as a sign of cirrhosis.

In addition, wedged hepatic venography investigates only a very small part of the liver where the lesions are not necessarily the same as in other places.

This is probably why in cirrhotic patients with a reversal of the portal flow some discrepancies were observed between the results yielded by arteriography and hepatic venography: the reversed portal flow demonstrated by selective hepatic artery injection could not be visualized by a wedged hepatic venography performed 2 or 3 days later (18).

Although hepatic venography has been widely used in cirrhosis it seems useful in two main cases: either to assess hepatic vein obstruction in Budd-Chiari syndrome, or to demonstrate the pre- or post-sinusoidal site of an intrahepatic obstacle.

III. Arterioportography

Arterioportography allows for a more complete and more physiological evaluation of the liver circulation, giving information about the liver morphology, the whole portal system and the portal flow, the hepatic arterial flow, the spleen and the splenic circulation and even in some cases on the patency of hepatic veins (15, 19, 20, 21, 22, 23).

The portal vein evaluation relies essentially on

the venous phase of superior mesenteric arteriography as the mesenteric vein flow rate is much less hindered as the splenic venous outflow in portal hypertension.

Injection in the superior mensenteric artery of 60 to 70 ml of a low viscosity contrast medium with a 6 ml/sec flow rate allows in most cases for a good opacification of the mesenterico-portal axis (22–25). The better portal vein visualization is obtained from 14 to 18 seconds after the beginning of the injection. Then it decreases slowly, resulting in a dense opacification of the liver parenchyma (portal hepatography), well demonstrated in normal persons at the 22nd second (Fig. 3).

The hepatic artery is studied by coeliac arteriography (45 ml of contrast medium at 10 ml/sec flow rate) or selective hepatic arteriography (30 ml of contrast medium at a 7 ml/sec flow rate). On the late films in normal persons only a faint arterial hepatography is observed because of the dilution of the contrast medium by the non-opacified portal blood.

The spleen and splenic vessels are well documented by coeliac arteriography. In normal conditions splenic and portal veins are fairly visualized between 7 and 12 seconds.

Usually, arterioportography gives no information about hepatic veins. In some cases, however, a hemodynamical abnormality is demonstrated, which is an indirect sign of obstruction of the hepa-

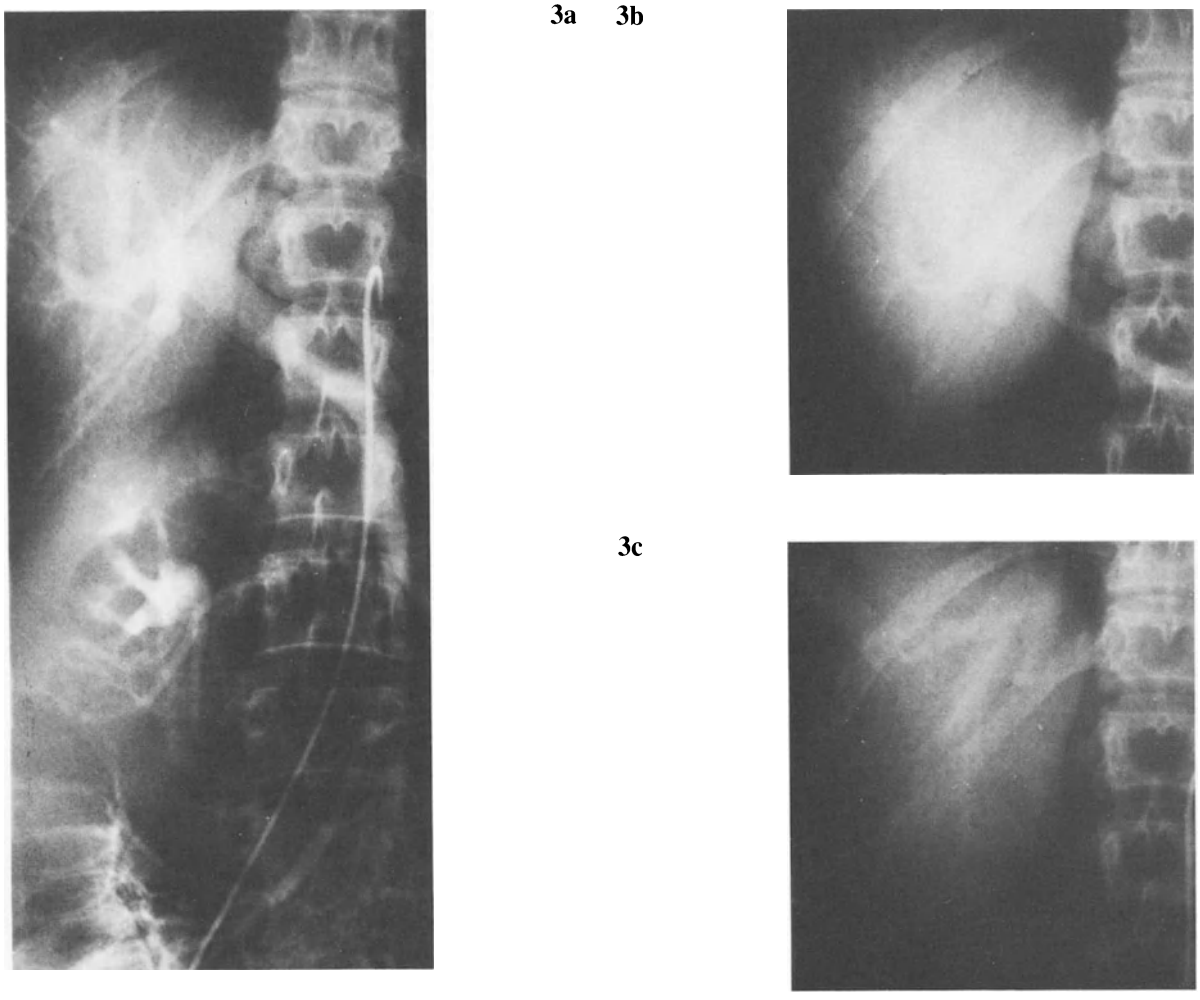


Fig. 3. Normal venous phase of superior mesenteric arteriography (S.M.A.).
3a: Normal mesenterico-portal axis is opacified 6 sec after the end of the injection of contrast medium.
3b: 6 sec later: normal portal hepatography.
3c: Hepatic vein opacification seen 3 sec later is an inconstant finding.

tic veins, the hepatic artery portal vein shunting resulting in the reversal of portal vein blood flow within the liver.

IV. Computed tomography

Owing to injection of contrast medium, C.T. may provide interesting information about the liver blood flow (26). The film taken 15 seconds after a bolus injection of contrast medium, in an upper limb vein corresponding to the arterial phase; the hepatic artery can be seen at the level of liver hilum but its intrahepatic branches are too thin to be visualized in normal persons. A second film taken at the 30th second corresponds to the venous phase and demonstrates a dense opacification of the portal vein and its intrahepatic branches. The contrast medium begins to accumulate within the hepatic parenchyma. The denser hepatogram is visualized on a third film at the 45th second (about 95 Hounsfield unit).

Angioscan has proved to be useful in demonstrating some hemodynamical abnormalities localized in a lobe or a segment of the liver. These abnormal angioscan pictures could be misinterpreted as morphological changes but the comparison with arterioportography permits their hemodynamical significance to be clearly understood. In addition, some vascular abnormalities such as collateral pathways or portal vein thrombosis can sometimes be demonstrated by angio-C.T.

3. THE MAIN ARTERIOGRAPHIC SIGNS OF PORTAL HYPERTENSION AND THEIR HEMODYNAMICAL SIGNIFICANCE

Three main angiographic elements permit the obstruction to the portal inflow to be recognized as well as its degree and its progression rate to be evaluated. These are:

- The quality of portal opacification on the venous phase of superior mesenteric arteriography;
- The spleen pattern;
- The hepatic artery changes.

I. The mesenterico-portal axis opacification

In case of intra- or sus-hepatic obstruction, the venous phase of superior mesenteric arteriography only allows for a reliable evaluation of the portal system. As a matter of fact, the venous phase of splenic arteriography has little interest from a hemodynamical point of view, as the splenic venous outflow decreases, then stops very early during the course of portal hypertension, whereas the mesenteric venous flow is maintained (Fig. 4).

From a physiological as well as a hemodynamical point of view, the superior mesenteric vein which carries to the liver the products resulting from intestinal absorption is truly the main branch of the portal vein, whereas the splenic vein seems to be a less important collateral vein. Therefore, 'splenoportal venous axis' is probably an inadequate though commonly used term, as such an axis does not exist from a physiological point of view, the real afferent venous way to the liver being the mesenterico-portal axis.

A faint opacification of the portal vein on the venous phase of coeliac arteriography is then often one of the first angiographic signs of portal hypertension; the more severe the obstruction, the slower the splenic venous flow, which can be interrupted and even reversed. On the contrary, the venous phase of superior mesenteric arteriography permits the relative value of the hepatopetal flow to be evaluated as compared to the hepatofugal flow according to the size of the vessels and the density of the opacification in each territory.

At an early stage of the disease, the normal portal hepatography is no more visualized, then fewer portal branches are seen within the liver and their size decreases whereas the porto-caval hepatofugal flow increases. However, the portal opacification permits only an approximate evaluation of the decrease of hepatopetal portal flow. In addition, this evaluation is only possible in case of intra- and supra-hepatic obstruction in which the portal vein is patent.

Thus, two other groups of angiographic signs, the spleen pattern and the evaluation of the arterial blood supply to the liver, seem to be much more useful signs. As a matter of fact they are the two main arteriographic signs of portal hypertension because their evaluation is always possible in every case of obstruction whatever its site and cause.

II. The spleen pattern

Splenomegaly is a classic sign of portal hypertension but its significance is not yet clearly understood, so it is often misinterpreted from a diagnostic and physiopathological point of view. A moderate splenomegaly may not be detected by clinical examination and may remain unknown for a long time. On the contrary, a very large splenomegaly, often associated with hypersplenism, may more easily be wrongly related to a hematological disease if the portal hypertension is inconspicuous, or be considered as the primary cause of the disease, responsible for the portal hypertension. In fact, the splenomegaly seems to be only the result of the venous stasis upstream of the obstruction and its size depends mainly on the degree and the evolution of this obstruction.

1. Angiographic signs

The increased size of the spleen is often the first sign of portal hypertension. When enlargement of the spleen is moderate as it is the case in the early stage of the disease, it may not be detected by clinical examination and is often demonstrated only by arteriography (Fig. 5).

The measurement of the long axis of the spleen at the splenographic phase seems to be a sufficiently reliable criterion to estimate the spleen size: it is normally shorter than 14 cm. In fact, there are relatively large individual variations (7 to 13 cm in our experience) so that it is not a very sensitive criterion. On the contrary, a long axis of the spleen greater than 14 cm is a reliable sign of a splenomegaly.

In addition, it seems usuful to appreciate the modifications of the splenic vessels and particularly of the splenic artery whose size (the mean normal diameter is 7 mm) increases together with the spleen size. In the same way, the more widened the artery, the more lengthened it is. Consequently, when the long axis of the spleen is no greater than 14 cm, widening and lenthening of the splenic artery may be valuable and helpful indirect signs to account for a moderate splenomegaly.

The splenic venous stasis results essentially in a faint opacification of splenic and portal veins on the venous phase of coeliac arteriography in spite of a very dense, homogeneous and more or less long-standing splenography. At the first stage of the disease, the splenic vein is well opacified but the portal vein and its intrahepatic branches are only faintly visualized because of the dilution of the contrast medium by the non-opacified mesenteric venous blood. In a second stage, the visualization of the splenic vein itself is faint and delayed; then in case of severe portal hypertension, the splenic vein is never visualized. The splenic vein size is variable because it depends on the splenic size as well as on the venous outflow rate. However, the fact that the narrower the splenic vein the slower its blood flow rate does not seem questionable.

Finally, splenic stasis may influence the splenic arterial circulation itself, giving rise to a slower splenic arterial flow which is sometimes clearly apparent on the films taken at different times during arteriography. In most cases, however, impairment of the splenic arterial flow results in an indirect sign, which is the decrease of the caliber of splenic artery and its intrasplenic branches which become abnormally thin (Figs. 6 & 7).

In some cases and probably after a long duration of the disease, lesions of the arterial wall may occur giving rise to aneurysms or medial fibrosis, which are probably related to the hemodynamical disturbance. These angiographic signs may be associated in a variable way, depending on the degree and the progression rate of the obstruction, whatever its cause.

2. The four main angiographic patterns of the spleen

a) Minimal splenomegaly without apparent hemodynamical abnomality of splenic circulation

When the splenomegaly is minimal, the long axis of the spleen is slightly increased (14 to 18 cm) but the spleen shape remains normal. Intra-splenic vessels are regularly disposed and a dense and homogeneous splenography is visualized. The splenic artery and its branches are slightly dilated. The splenic vein is well opacified and sometimes slightly widened as well as the artery. A good opacification of the portal vein and its main branches within the liver is visualized: the venous phase of coeliac arteriography is normal.

That is the pattern of a congestive splenomegaly due to a moderate splenic stasis without an important decrease of the splenic venous outflow. It may be due to:

- either a minimal and recent obstacle related for instance to a posthepatitis cirrhosis. It is, however, an uncommonly encountered case as this minimal splenic stasis is usually clinically unapparent (Fig. 5). It corresponds to the early stage of the portal hypertension which remains usually unrecognized,
- or a more severe obstruction but perfectly compensated by the development of an important collateral flow so that portal hypertension is minimal and remains clinically unapparent for a long time (Fig. 33, 35, 36).

b) Moderate splenomegaly with splenic venous stasis

A moderate splenomegaly is clearly apparent, the

spleen becoming round shaped with a long axis ranging from 16 to 20 cm. The splenic artery is dilated and lengthened but the diameter of its intrasplenic branches, which are more or less stretched, remain normal. The splenic vein diameter remains normal and is not as widened as it should be according to the spleen enlargement. The decreased splenic venous outflow results in a poor opacification on the venous phase of coeliac arteriography.

The portal vein and its intrahepatic branches are faintly visualized, whereas a good opacification of the splenic vein remains in some cases. Then a faint and delayed opacification of the splenic vein is seen while its diameter decreases and the hepatofugal collateral flow increases.

This is the common pattern of the decreased hepatopetal portal flow, due to a moderate and more or less quickly progressive obstruction, resulting in a usually moderate portal hypertension. The spleen becomes more enlarged as the disease responsible for the obstruction is more slowly progressive. This angiographic spleen pattern is commonly encountered in alcoholic cirrhosis as well as in cases of complete portal vein obstructions, with too few bridging collaterals to preserve hepatopetal flow (Fig. 23).

c) Minimal splenomegaly with slowed arterial splenic flow

In some cases the hepatopetal splenic venous outflow is very much decreased or even completely interrupted. The splenic vein is narrowed and very faintly visualized on the delayed films only. In some cases, no opacification of the splenic vein can be achieved. This interrupted splenic venous outflow corresponds to a functional obstruction pattern, due to high and rapid rise of portal pressure, resulting in an impairment of the arterial flow itself.

The slowing of splenic arterial flow gives rise to a narrowing of the splenic artery better visualized on its intrasplenic branches which are abnormally thinned whereas the splenography is delayed and prolonged. In rare cases the delayed opacification of the splenic artery and its branches is clearly apparent on the successive films of the coeliac arteriography as compared to hepatic arterial opacification (Figs. 6–9). In most cases, however, the thinning of intrasplenic arterial branches is the best sign of this 'low flow rate pattern' of the spleen.

The same angiographic pattern of arterial vasoconstriction may be seen everywhere in the body whenever an acute obstruction of the venous outflow tract occurs and eventually results in ischemic lesions. Arterial vasoconstriction has been well demonstrated to occur for instance in the femoral artery as a result of 'Phlegmatia coerulaea' as well as in the mesenteric artery in case of mesenteric ischemia due to mesenteric vein thrombosis (27, 28).

It is easy to understand that a markedly reduced splenic flow rate hinders the splenomegaly to occur, so that this 'low flow rate pattern' is always associated with a normal-sized or near normal-sized spleen. Similarly, atherosclerotic stenoses of the splenic artery results in a decreased spleen size as commonly seen in elderly patients.

In the course of portal hypertension it is commonly found that the more reduced the splenic flow rate, i.e. the more severe the portal hypertension, the smaller the splenomegaly. This angiographic spleen pattern is usually the result of a very tight, recent and quickly progressive obstruction to the hepatopetal portal flow, resulting in a quick rise of the portal pressure. In this condition the splenic arterial flow is markedly slowed before the spleen can enlarge.

Budd-Chiari syndrome is the commonest clinical condition to determine this angiographic pattern (Fig. 10). It can also occur in some case of quickly progressive alcoholic cirrhosis associated with an unusually poor hepotafugal collateral flow. It is less common in prehepatic obstructions (Fig. 29).

d) Very large splenomegaly

Splenomegaly may be very large with a maximum height greater than 20 cm, occupying the whole left hypocondrium and producing a marked displacement of the stomach and bowel as well as the left kidney. Splenic artery is markedly widened, lengthened and tortuous, and its branches, more or less stretched according to splenic enlargment, have a regular course within the spleen. The splenography is dense and homogeneous and the splenic vein is widened as well as the portal vein.

The venous phase of splenic arteriography is usually poor, the splenic vein being faintly opacified because of the dilution of the contrast medium

within this huge splenomegaly. Of course the more decreased the splenic venous flow, the poorer the splenic vein opacification, but portal blood stasis and contrast medium dilution within the enlarged spleen have the same result, so that it is very difficult to evaluate if the splenic venous outflow is normal or not when the spleen is so much enlarged.

This angiographic spleen pattern seems to be the result of a long-standing moderate and slowly progressive obstruction to the hepatopetal portal flow, whatever its cause (Figs. 11, 12, 13, 14, 15). Indeed, as it takes a long time for such a large splenomegaly to occur, the disease responsible for the obstruction to the portal inflow must be a very slowly progressive one. In addition, the splenic circulation should not be too much slowed as a result of the decreased portal flow. For this reason the obstruction must be a moderate one.

A very large spleen is thus the result of a mild, long-standing and slowly progressive portal hypertension. It may be the only sign of the disease, often associated with hypersplenism. It is therefore sometimes misinterpreted as the result of a hematological disease. In addition, the portal pressure is only slightly increased, no higher than 15 to 18 mm Hg, in some cases so that portal hypertension may be thought questionable.

These hemodynamical conditions may be fulfilled in many clinical circumstances such as:

- hepatic fibrosis from unknown origin as well as some cases of post-hepatitic cirrhosis (Fig. 11);
- the rare cases of congenital obstruction of the hepatic veins;
- congenital hepatic fibrosis or cirrhosis (hemochromatosis, Wilson's disease) (Fig. 13);
- every case of moderate obstruction where the disease process progression has been stopped or slowed either spontaneously or after medical treatment or surgical portocaval anastomosis. For instance, some cases of long-standing alcoholic cirrhosis (Fig. 14) or Budd-Chiari syndrome may be associated with a pronounced splenomegaly (Fig. 18).

e) Greatly enlarged spleen associated with splenic arterial wall lesions

The relative frequency of splenic artery aneurysms in the course of portal hypertension has been demonstrated by Boijsen (29) and Williams (30) who stated that they occurred in 15% and 31% of the cases, respectively.

The splenic aneurysms are much more frequent as the splenomegaly is more pronounced, that is to say, the angiographic pattern suggests a long-standing and slowly progressive portal hypertension. According to Williams, splenic artery aneurysms frequency could be as high as 43% in a group of patients with greatly enlarged spleens.

This frequency of splenic artery aneurysms in long-standing portal hypertension associated with a pronounced splenomegaly is widely confirmed by our own experience. In some cases, the aneurysms which did not exist at an early stage of the disease were shown to appear a few years later whilst the spleen had greatly enlarged (Fig. 10).

In most cases they are small and often multiple aneurysms distally located at the bifurcations of intrasplenic arterial branches. In some cases, the aneurysm is located on the splenic artery itself. Its size is usually moderate (1 to 3 cm) and no clinical symptom can be referred to this lesion which is but an angiographic discovery (Figs. 15, 16, 59, 63). However, a complication is theoretically possible and in our experience, it occurred in two cases. In one case of alcoholic cirrhosis the splenic artery aneurysm ruptured into the peritoneal cavity giving rise to hemoperitoneum and was discovered by emergency arteriography. In another case of congenital Budd-Chiari syndrome, large-sized splenic artery aneurysms were shown to have ruptured into the splenic vein, producing a splenic arteriovenous fistula (Fig. 18). Thus, the fact that portal hypertension may influence the development of splenic artery aneurysms, mainly in long-standing and slowly progressive forms of the disease seems unquestionable.

The arterial wall abnormalities probably result from either the widening and the lengthening of the splenic artery, which are always associated with the spleen enlargement or the hemodynamical changes of the splenic blood flow. Indeed, aneurysms are well known to occur as the late result of arterial widening associated with increased flow rate elsewhere in the body: for instance, aortic coarctation resulting in intercostal arteries aneurysms is another example of this phenomenon.

As few cases have been submitted to histological examination, these arterial wall lesions are not very

well known from a pathological point of view. In one of our cases where a splenectomy was performed, pathological examination disclosed that the aneurysmal wall as well as the splenic arterial wall were involved by lesions of medial fibrous dysplasia, whereas there were no arterio-sclerotic lesions (31, 32).

f) Splenomegaly and splenic artery fibro-muscular dysplasia

Although very rare, an irregular splenic arterial wall looking like the 'string of beads' angiographic pattern of the fibro-muscular dysplasia was found out in 3 or 4 of our cases of portal hypertension associated with a greatly enlarged spleen. One of these patients underwent splenectomy and on histological examination the splenic artery was shown to be involved by the same medial fibrosis as described in arterial fibro-muscular dysplasia (Fig. 17).

The question arose as to whether this splenic medial fibrosis could be an acquired lesion of the arterial wall probably induced by the haemodynamic disorder as suggested for splenic artery aneurysms. In order to answer this question, fibrous replacement of the medial arterial layer was searched for systematically by histologic examination of the splenic artery in 37 patients who underwent surgical splenorenal shunts or splenectomy for portal hypertension.

The pathological examination of the excised splenic arteries was carried out in many samples with special tainting. Thirteen out of the 37 patients were shown to have medial fibrosis. In every case the arterial wall exhibited an irregular thickness due to alternating zones of medial fibrous hyperplasia and destruction (the latter representing mural aneurysms) and alterations of the internal elastic lamina which was thick and reduplicated or focally absent. Other layers were normal and no sign of atheroma was found (31, 32) (Figs. 15, 61, 75).

Although surprising, these results were confirmed by several pathologists who agreed with the fact that the histologic pattern was exactly the same as described in the medial fibroplasia, which is by far the most common form of fibromuscular dysplasia, as described elsewhere in the body and mainly in the renal arteries.

Only one out of these 13 patients had the typical 'string of beads' angiographic appearance of the splenic artery. In the other 12 patients the splenic artery was only dilated and lengthened as in every case where the spleen is greatly inlarged, but four of them had also a splenic artery aneurysm (Fig. 15 and 75) (1).

Such a frequency (35%) of this splenic artery medial fibrosis associated with a long-standing portal hypertension is quite unexpected. It may be compared to the frequency of splenic artery aneurysms already described in the same group of patients (29, 30). It is then strongly suggested that this medial fibrosis may be related to the splenic morphological and hemodynamical changes resulting from a long-standing portal hypertension.

As these lesions compare exactly with those of the fibro-muscular dysplasia the question is raised as to whether it may be the same disease or not. In the first case, one might wonder if the arterial fibro-muscular dysplasia is not at least in some cases the result of a hemodynamical disturbance such as increased resistance to the blood flow or chronic vasoconstriction, which would be of great interest from a physiopathological point of view.

In conclusion, in the course of the obstructions to the hepatopetal portal flow, the spleen changes seem to be essentially related to portal hypertension, depending on its degree and its evolution. Enlargement of the spleen is all the less as portal hypertension is higher and more quickly progressive.

Thus, the splenic changes due to portal hypertension may be compared to the urinary tract changes related to ureteral obstructions: acute and severe ureteral obstructions (such as calculous or carcinomatous ones) do not result in a pronounced ureteral dilatation above the obstruction as dramatic pressure increase gives rise to an impairement of the renal urinary secretion which is greatly decreased.

On the contrary, in cases of moderate and slowly progressive obstruction (such as megaureter or uretero-pelvic junction syndrome) a very large dilatation of the urinary tract always occurs above the obstruction. In the same way, in mitral stenosis, a tight stenosis is often associated with little dilatation of the left atrium and a severe venous pulmonary hypertension occurs. On the other hand, a

mild or moderate mitral stenosis may be associated with a great enlargement of the left atrium while the venous pulmonary hypertension is less severe. Hence, obstructions to the hepatopetal portal flow are probably associated or not with a greatly enlarged spleen in the same way as there are urinary obstructions with or without great dilatation of the urinary tract and mitra stenosis with or without a large left atrium. Theoretically, other parameters than portal hypertension may of course influence the spleen enlargement.

When associated with portal hypertension, infectious or parasitic diseases, as well as haematological and reticulo-endothelial system diseases may contribute to splenomegaly. On the contrary, splenic artery arteriosclerosis in elderly patients may result in a decreased spleen size or hinder a portal hypertension to result in splenomegaly.

In practice, however, these cases seem too rare for the value of splenic changes as a witness of portal hypertension to be reduced. The angiographic spleen pattern truly appears to be essentially related in most cases to the degree and the progression rate of portal hypertension.

III. Hepatic arterial changes

In most patients with moderate to severe portal hypertension the hepatic artery and its branches become dilated, which is the result of an increased hepatic arterial flow. Indeed, arteries dilate to accommodate an increased flow. Furthermore, this increased arterial flow is well demonstrated by two other associated angiographic signs: the reversed flow through the gastro-duodenal artery which contributes to feed the liver from the superior mesenteric artery and the denser arterial hepatography as normally visualized on the late films of hepatic arteriography.

However, it may be difficult to recognize this hepatic artery widening when it is minimal. The common hepatic artery diameter can be measured on the films: it is normally no greater than 7 mm in most cases. But this measurement is not a reliable criterion because of individual variations and frequency of multiple hepatic arteries.

However, hepatic artery dilatation is better appreciated on the intrahepatic branches than on the arterial stem itself, because the hepatic arterial ramifications are usually thin in normal conditions mainly at the periphery of the liver. Hence, a dilatation with elongation tortuosity and sometimes a duplicated appearance of the peripheral arterial branches within the liver associated with a dense arterial hepatography are the two main angiographic signs suggesting an increased arterial flow to the liver. The angiographic evaluation of this increased arterial flow is very interesting from a hemodynamical point of view as it results from a decrease of the hepatopetal portal flow.

Although it may be difficult to demonstrate unequivocally this point by flow rate measurements, most physiological and angiographic studies permit to estimate that there exists a close correlation between decreased portal inflow and increased hepatic arterial flow which is frequently and widely admitted. Because communications exist within the liver between arterial branches and portal venules either through the sinusoids, or probably also through presinusoidal shunts, it is likely that the hepatic arterial flow increases to compensate for a decreased portal inflow.

A presinusoidal portal obstruction will result in a decreased pressure in the downstream located portal veins as well as in sinusoids. The pressure gradient between these veins and the hepatic arteries is then increased, which is likely to determine an increased arterial flow.

In case of post-sinusoidal obstruction the increased intrahepatic resistance can probably give rise to a severe impairment of the hepatopetal portal flow because of the normally low pressure gradient between portal veins and hepatic veins. Because arterial pressure is much higher than portal pressure, the hemodynamical conditions become more favorable to the arterial flow as compared to the portal flow.

By measuring arterial and portal flow rates during surgery in some patients, Schenk was able to prove that the hepatic arterial inflow increases to compensate for the lost portal flow. When the portal flow was interrupted, he found that the arterial flow increased to 80% in patients with normal liver and up to 400% in cirrhotic patients (33).

This relationship between a decreased portal inflow and an increased hepatic arterial flow has been supported for years by arterioportography,

although this method allows only for a relative evaluation of the arterial and portal flow rates as compared either to each other or to the normal angiographic pattern. However, in some rare cases where the portal obstruction either involves only some intrahepatic branches while other remain patent or varies with time in the same patient, the arterial changes documented on arterioportography may be particularly demonstrative (Figs. 19 & 20.

In case of partial, intrahepatic portal vein thrombosis resulting from an acute pancreatitis, for instance, arterioportography demonstrates that intrahepatic arterial branches dilate in the segments where the portal branches are obstructed, while they remain normally thin in the segments where portal branches remain patent. These arterial changes are clearly apparent when the diameters of arterial branches are compared to each other. However, the increased arterial flow is better demonstrated on the late films where an abnormally dense arterial hepatography is visualized only in the liver segments where portal veins are obstructed. In the other segments where portal veins are patent, only portal hepatography is seen on the venous phase of superior mesenteric arteriography as usually in normal persons.

In this case, arterial and portal hepatography result in a 'mirror image', clearly demonstrating that the hepatic arterial inflow increases to compensate for the lost-portal flow, the parts of the liver parenchyma no longer fed by portal vein flow being supplied by arterial blood (Fig. 19). Comparison of two arterioportographies performed in the same patient at different stages of the disease also demonstrates the close relationship between hepatic arterial changes and the variations of the hepatopetal portal flow rate.

It is the case, for instance, in cirrhotic patients treated by surgical porto-caval anastomosis whose hepatopetal portal flow is interrupted and even sometimes reversed, the portal vein becoming an outflow tract.As a result of this a pronounced dilatation of the hepatic artery is seen to occur in every case, as compared to the preoperative study. In addition, it is suggested from our experience that the higher the hepatofugal portal flow the more dilated the hepatic artery (18).

On the other hand, if an obstruction to the hepatopetal portal flow improves so that the portal flow rate increases, the hepatic arterial dilatation is shown to reduce by arterioportography as compared to a previous study. This hemodynamical condition may be encountered, for instance, in case of portal thrombosis where arterioportography performed at an early stage of the disease shows a greatly decreased portal inflow because the hepatopetal collateral network has not yet developed. At that stage an important dilatation of the hepatic artery is demonstrated.

When arterioportography is repeated a few months later, bridging collaterals running parallel, the obstructed portal vein may have developed (cavernous transformation) and a better, although never normal, hepatopetal portal flow can be demonstrated with filling of peripheral portal vein branches through numerous subcapsular vessels. As a result of this the hepatic artery is shown to be less dilated than on the previous arteriography (Fig. 20).

Finally, it is perhaps possible for the hepatic arterial flow to decrease under normal values in case of an abnormally high hepatopetal portal flow rate but this rare hemodynamical condition is more difficult to demonstrate. However, in an uncommon case of diffuse angiodysplasia of the colon in a 29-year-old man, where arteriography showed a pronounced dilatation of superior and inferior mesenteric arteries followed by early and massive portal venous opacification through large and numerous arterio-venous shunts, an unusually thin hepatic artery with a poor opacification of its peripheral branches suggested a decreased hepatic arterial flow. In this patient a two-fold increase of the cardiac output could be measured, which was obviously the result of an increased portal inflow. The liver was found to be normal at surgery as well as the portal pressure. Despite this high portal flow rate, there was no portal hypertension (Fig. 21).

This relationship between the arterial and portal flow rates also seems to be well demonstrated by studying the results of arterioportography in a great number of cirrhotic patients, whatever the direction of the portal flow. In a study on the spontaneous reversal of intrahepatic portal flow in cirrhosis the hepatic arterial dilatation was found to be well correlated to the degree of the hepatofugal portal flow (18). It must then be admitted that the

hepatic arterial dilatation is usually the result of a decreased hepatopetal portal flow whatever its cause and that there is, in most cases, a good correlation between the lost portal flow and the increased arterial flow.

These arterial changes occur in every case of presinusoidal obstruction and particularly in portal vein thrombosis as well as in most post-sinusoidal obstructions once the hepatopetal portal flow is sufficiently reduced; this is mainly the case in cirrhosis.

There is, however, one exception to this rule, the Budd-Chiari syndrome, because the quickly progressive hepatic vein thrombosis gives rise to a unique hemodynamical condition which can be compared to an ischaemia of the liver from venous origin. This is the reason why the hepatic arterial flow can not increase.

To our knowledge the Budd-Chiari syndrome is the one case where a dramatic decrease of the hepatopetal portal flow does not result in an increased arterial flow. This point ought to be stressed as it results in a very typical angiographic pattern which will be described later.

Of course, the portal flow rate is not the one parameter inducing hepatic arterial changes, which may result from many other causes. For instance, any hypervascular liver tumor, aneurysm or vascular malformation is associated with a dilated hepatic artery and an increased arterial flow but these lesions are easily recognized by arterioportography and do not raise any diagnostic problem.

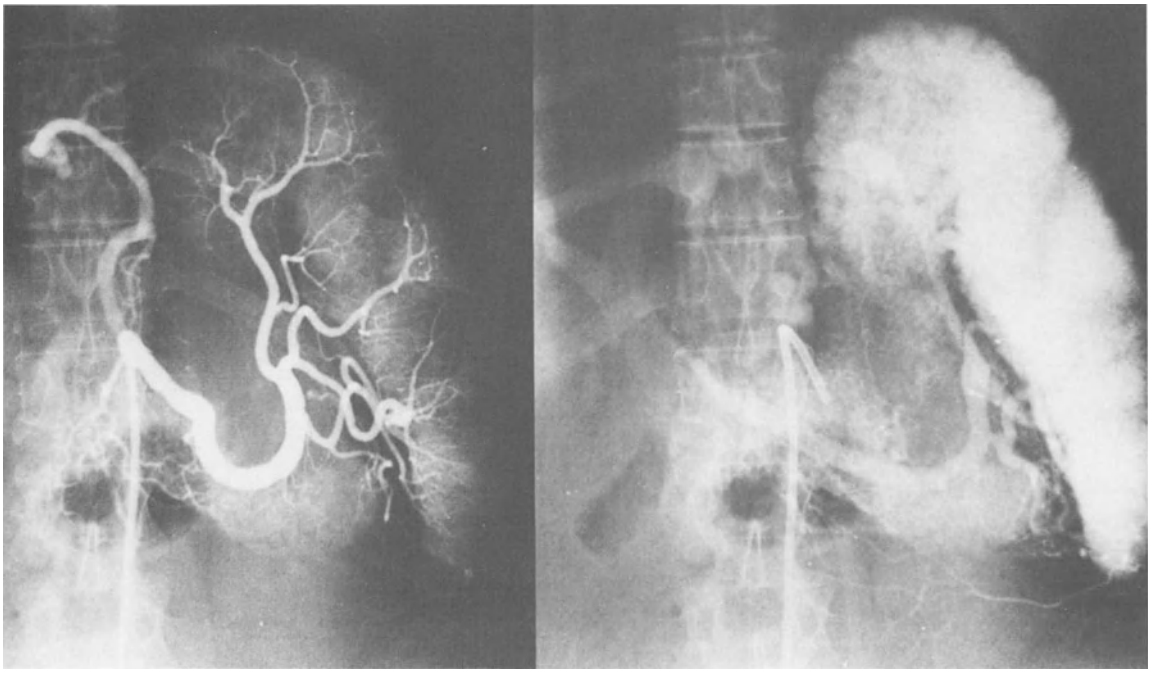
However, some diffuse liver diseases may be associated with a dilated hepatic artery whereas any specific angiographic sign permits to make the right diagnosis. It may be the case in virus infections, inflammatory diseases such as collagen or granulomatous diseases involving the liver, as well as the 'nephrogenic hepatic dysfunction syndrome' (34, 35, 36). These are, fortunately, rare clinical conditions, and the hepatic arterial widening is not associated with the other angiographic signs of portal hypertension.

Obstructive jaundice is usually associated with a pronounced hepatic arterial dilatation which is thought as resulting from a decreased portal inflow. Indeed the biliary stasis in the markedly dilated intrahepatic bile ducts could be responsible for a compression of intrahepatic portal veins as demonstrated by some experimental works (37). Thus hepatic artery embolisation in patients with obstructive jaundice can result in liver necrosis, as already reported. (38).

Most of these clinical circumstances being either infrequent or easily recognized the hepatic arterial changes may be considered in most cases as a good indirect witness of the hepatopetal portal flow rate. Hence the coeliac arteriography enables one to evaluate two important factors associated with any obstruction to the hepatopetal portal flow whatever its site and its cause but whose physiopathological significance is quite different:

- Hepatic arterial changes: indirect result of the hepatopetal portal flow decrease.
- Splenic changes: essentially related to the degree and the progressing rate of portal hypertension.

These two factors are usually associated as portal hypertension results from an obstruction to the portal inflow, but in some cases only one of these factors is present if either a long-standing portal hypertension is associated with a near normal hepatopetal portal flow or a severe obstruction is associated with a near normal portal pressure as in some cases where the development of very large porto-systemic collaterals occurs (Figs. 35, 36, 42, 49). In all cases a simultaneous evaluation of both angiographic abnormalities is at any rate necessary for the diagnosis and results in a better understanding of the disease in every case.

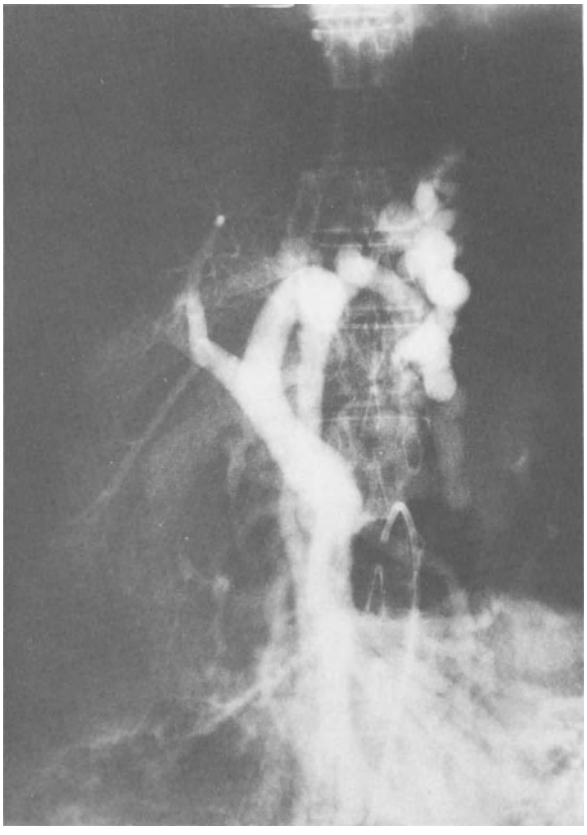


4a

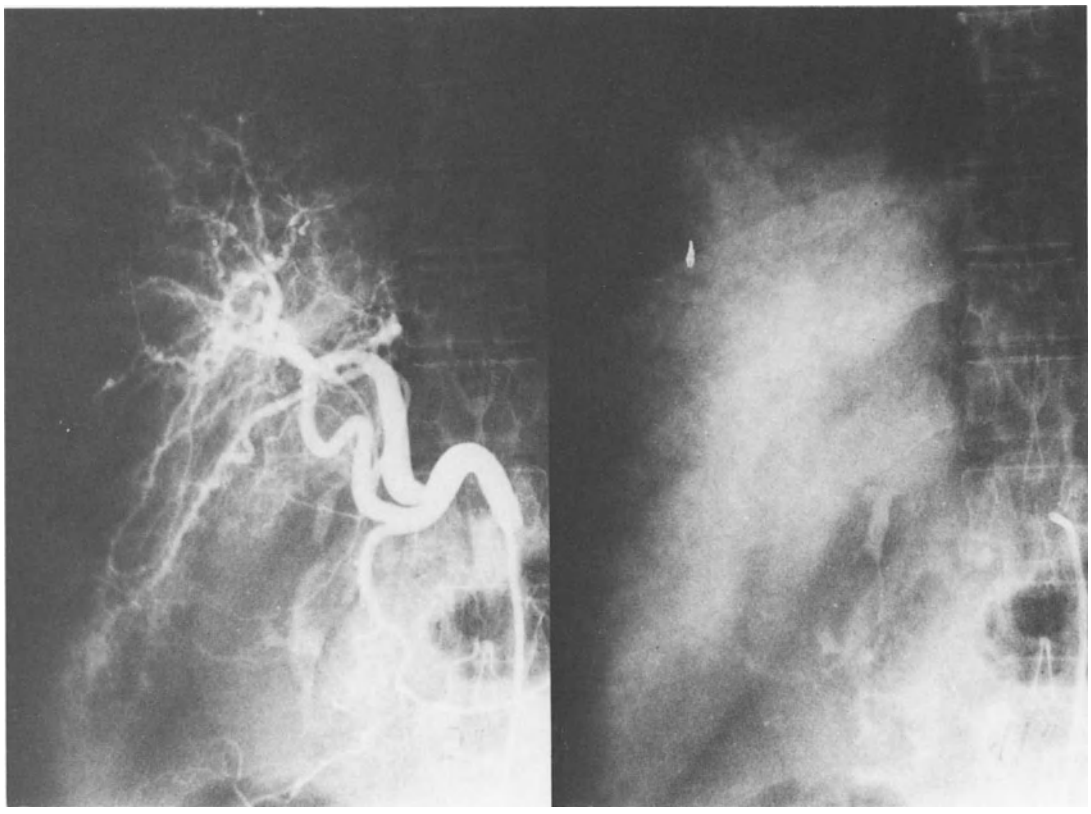
Fig. 4. Severe alcoholic cirrhosis in a 46-year-old female.

4a: Moderate splenomegaly (18 cm) with relatively thin intrasplenic arterial branches. Despite the selective splenic arterial injection resulting in a dense splenography the splenic vein is thin and poorly opacified, i.e., the splenic venous outflow is markedly reduced.

4b

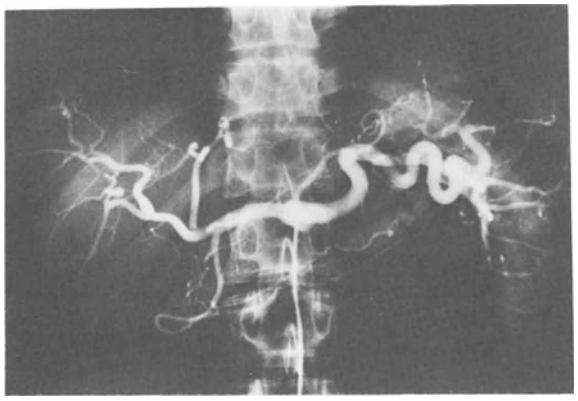


4b: However, on the venous phase of S.M.A., the mesenterico-portal axis is fairly well opacified, with hepatofugal flow through umbilical vein and gastric varices, whereas the narrowed and poorly opacified intrahepatic portal branches indicate a markedly reduced portal inflow.

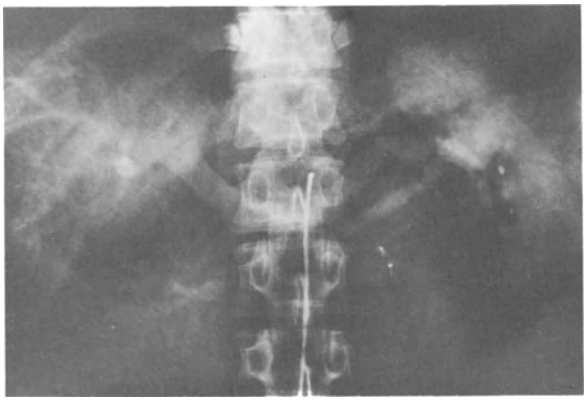


4c

4c: Owing to this the hepatic artery and its intrahepatic branches are markedly dilated and result in a dense arterial hepatography.



5a

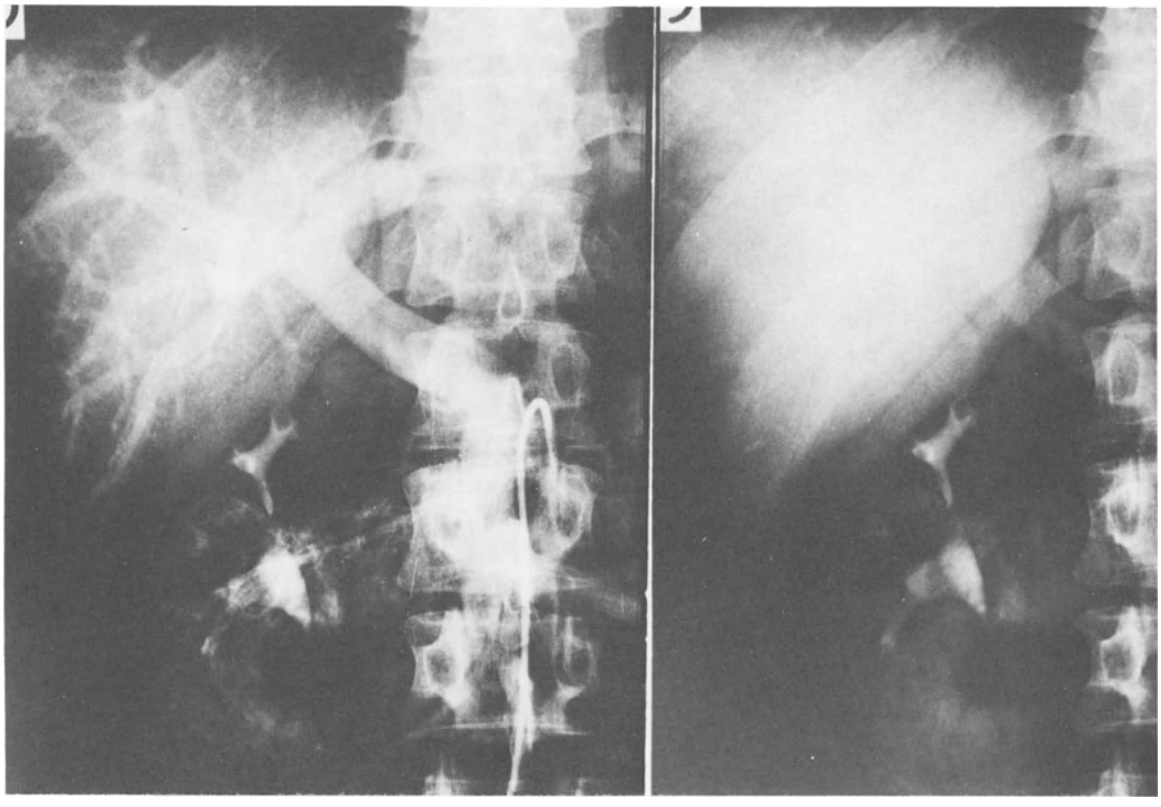


5b

Fig. 5. Chronic active hepatitis in a 34-year-old male (8/78). – virus hepatitis 6 months earlier (2/78): ALAT : 1.400 Ui – Hbs Ag +

5a: Normal hepatic artery. Moderate splenomegaly (17 cm). Widened and lengthened splenic artery.

5b: Normal splenography resulting in good opacification of splenic and portal vein.



5c

5c: Normal venous phase of the S.M.A.: intrahepatic portal branches as well as portal hepatography fairly well visualized. No hepatofugal flow.

Splenomegaly is the one sign of the incipiens portal hypertension. Virus hepatitis confirmed by laparoscopy and liver biopsy (moderate liver cell lesions without fibrosis).

One year later (8/79) checking liver biopsy shows more severe liver cell lesions and periportal fibrosis.

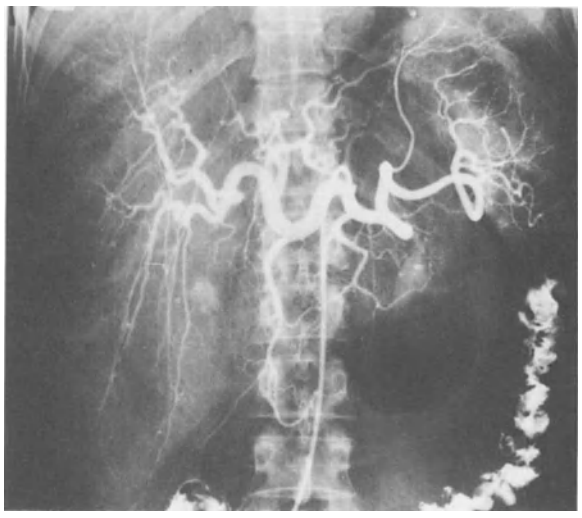
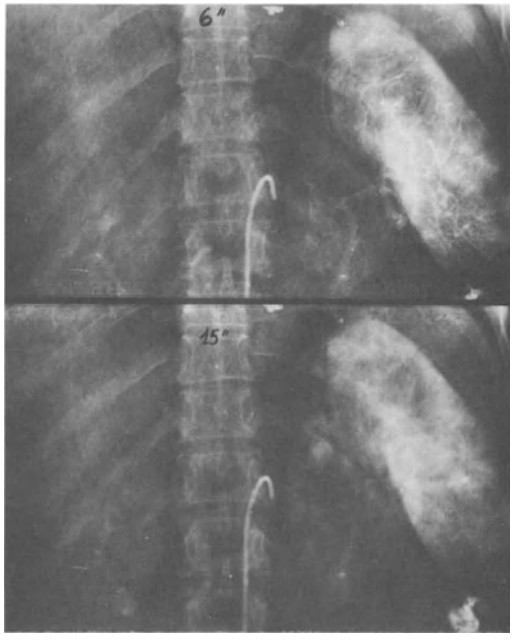


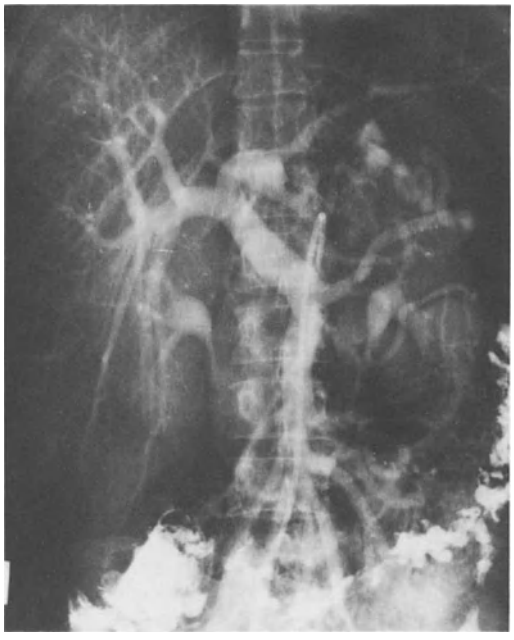
Fig. 6. 35-year-old female known to have alcoholic hypertrophic cirrhosis with ascitis for 6 months. Corrected sinusoidal pressure: 26 cm saline.

6a: Diffusely enlarged liver with a widened hepatic artery but only moderately dilated intrahepatic arterial branches. Minimal splenomegaly (15 cm) with very thin intrasplenic arterial branches.

6a



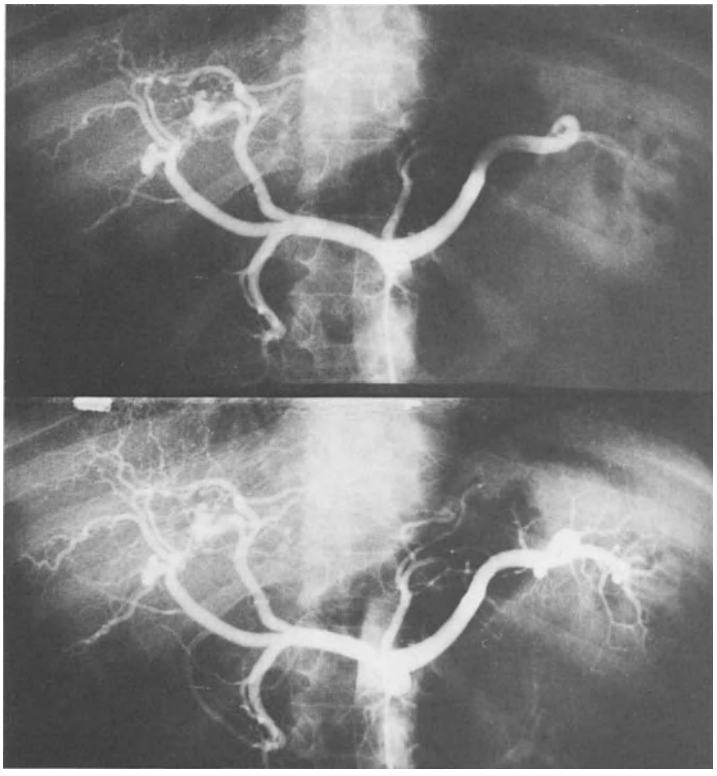
6b



6c

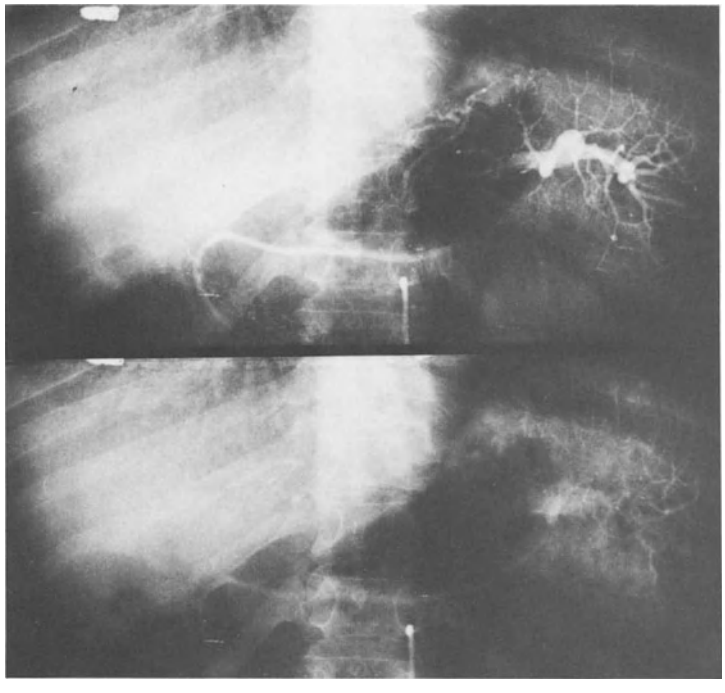
6b: Slowed splenic arterial flow clearly evidenced as compared to the hepatic arterial flow: persistent opacification of the intra splenic arterial branches at the 6th sec without visualization of the splenic vein.

6c: Moderately decreased mesenterico-portal flow to the liver with the tertiary portal vein branches still visualized but no portal hepatography. Hepatofugal flow through the splenic vein and short gastric veins thus explaining the functional obstruction of the splenic venous outflow.



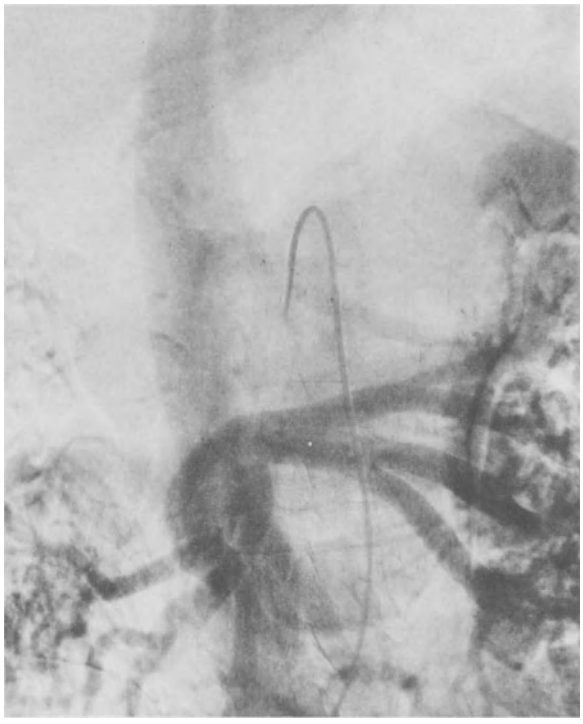
7a

Fig. 7. Alcoholic cirrhosis with bleeding esophageal varices in a 54-year-old male (23–3–77).
7a: Dilated hepatic artery accounting for a decreased hepato-petal portal flow. The slowed splenic arterial flow is well evidenced as compared to the hepatic arterial flow 1.5 and 4 seconds after the injection.



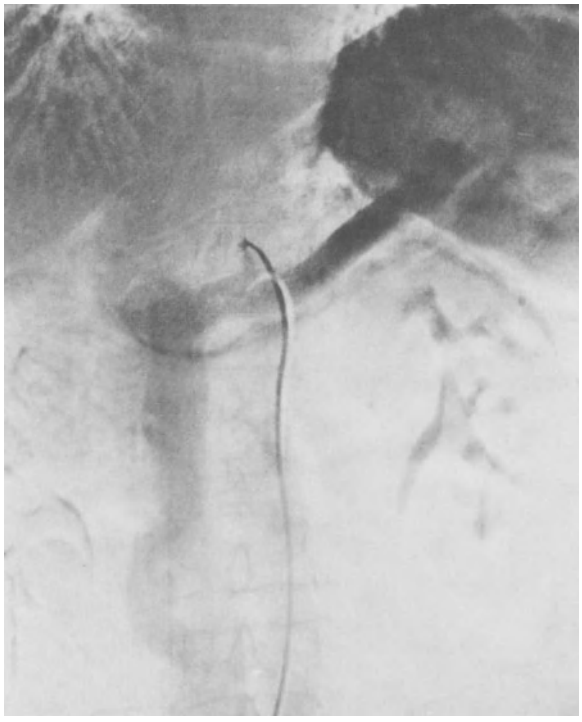
7b

7b: 6 and 12 seconds later, markedly thinned intrasplenic arterial branches remain visualized. Splenography is very poor and the splenic vein never opacifies.



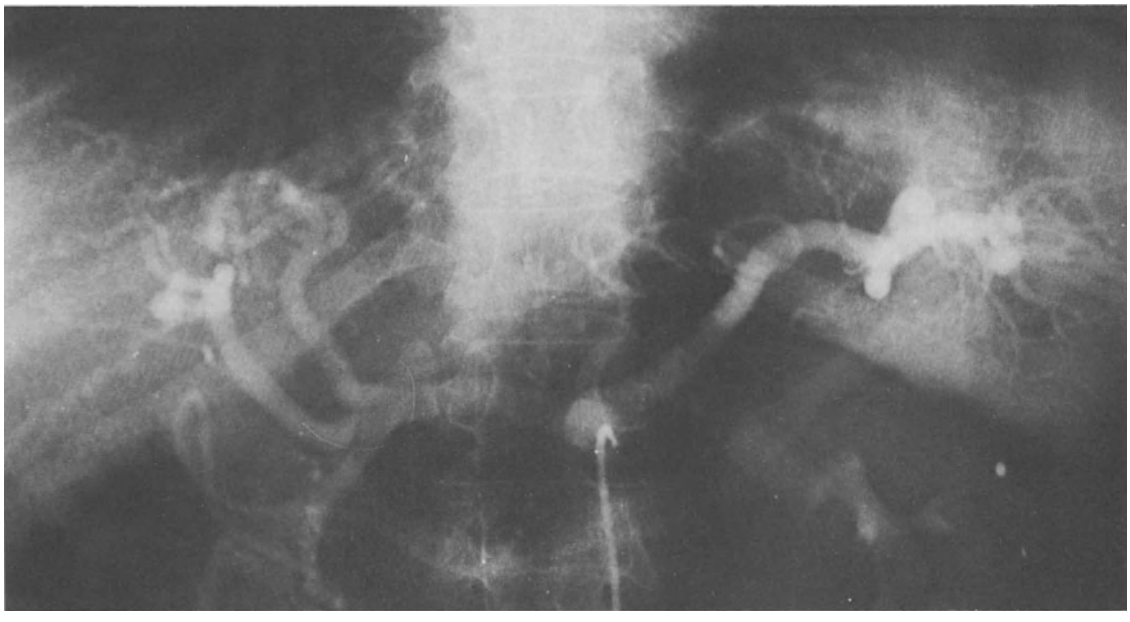
7c

7c: A meso-caval interposition shunt performed one month later is shown to be normally patent on the venous phase of S.M.A. (16–9–77).



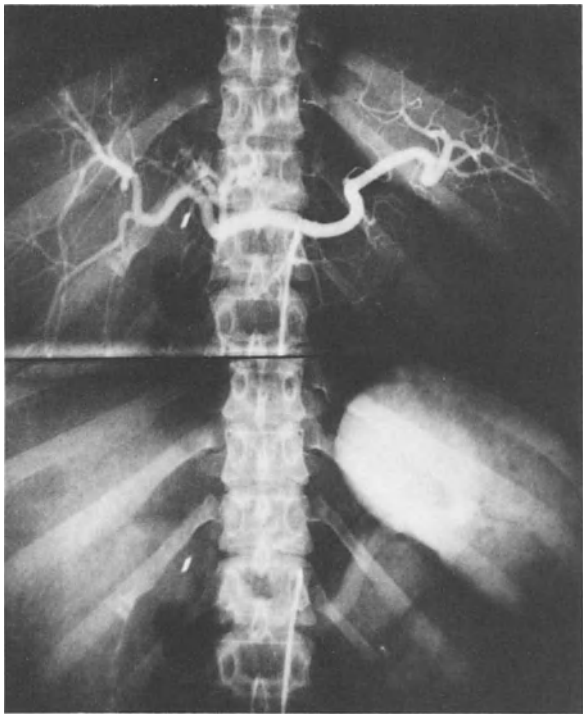
7e

7e: Since the splenic venous blood drains retrogradely through the superior mesenteric vein toward the graft and the inferior vena cava, together with the mesenteric blood.

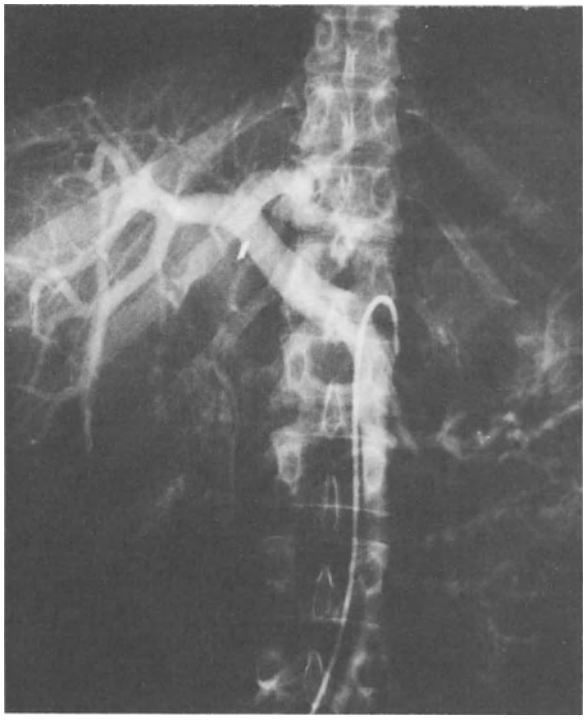


7d

7d: On coeliac arteriography (14–9–78) the splenic arterial flow is no longer slowed (Normal intrasplenic branches and early splenography).



8a

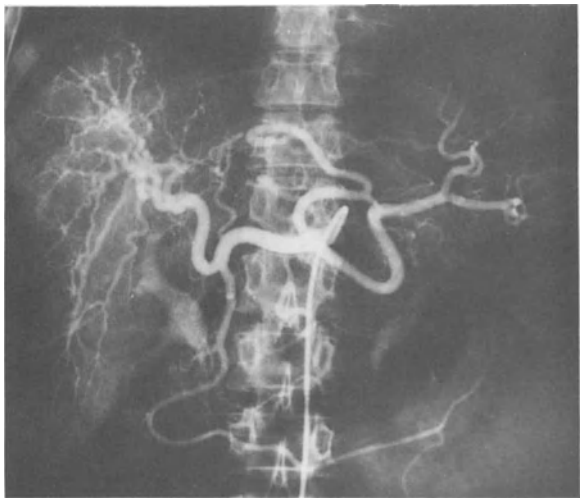


8b

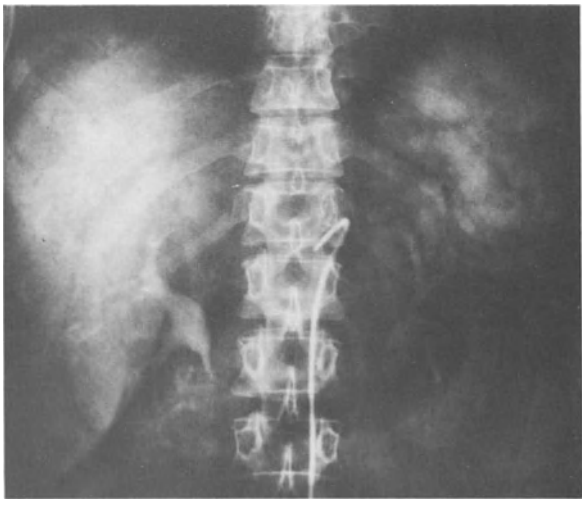
Fig. 8. Hypertrophic alcoholic cirrhosis in a 28-year-old female (3–5–78) intermittent jaundice, weight loss and weakness for 3 months.

8a: Moderately dilated hepatic artery
– Spleen size within normal limits (14 cm) with markedly thinned out intrasplenic arterial branches.
– Dense and prolonged splenography with only faint opacification of the splenic vein.

8b: Good portal vein filling on venous phase of S.M.A. with marked thinning of its intrahepatic tertiary branches and retrograde opacification of inferior mesenteric vein.
Angiographic pattern of a moderately decreased mesenterico-portal inflow associated with markedly reduced splenic venous and arterial flow resulting from a recent and rapidly progressive intrahepatic obstruction.



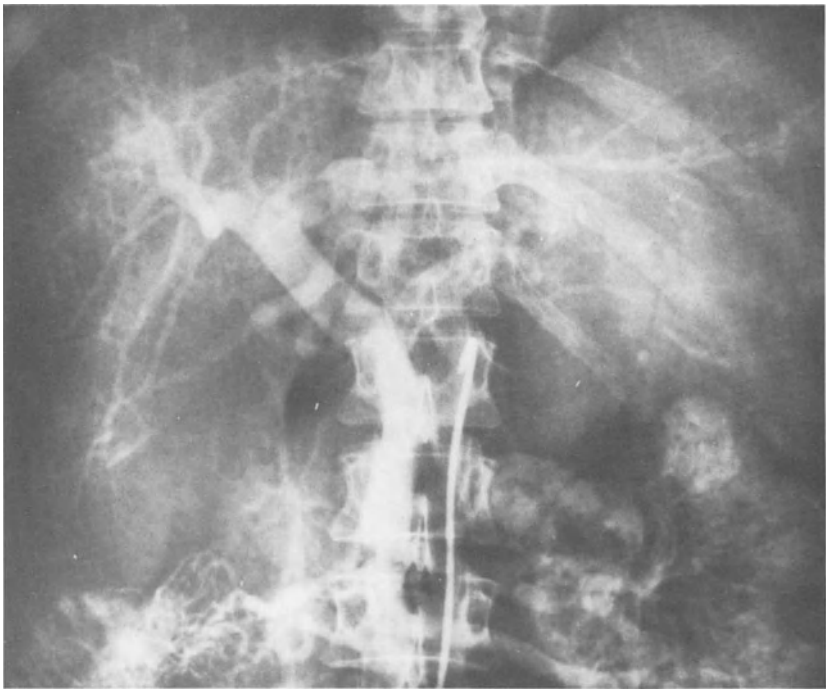
9a



9b

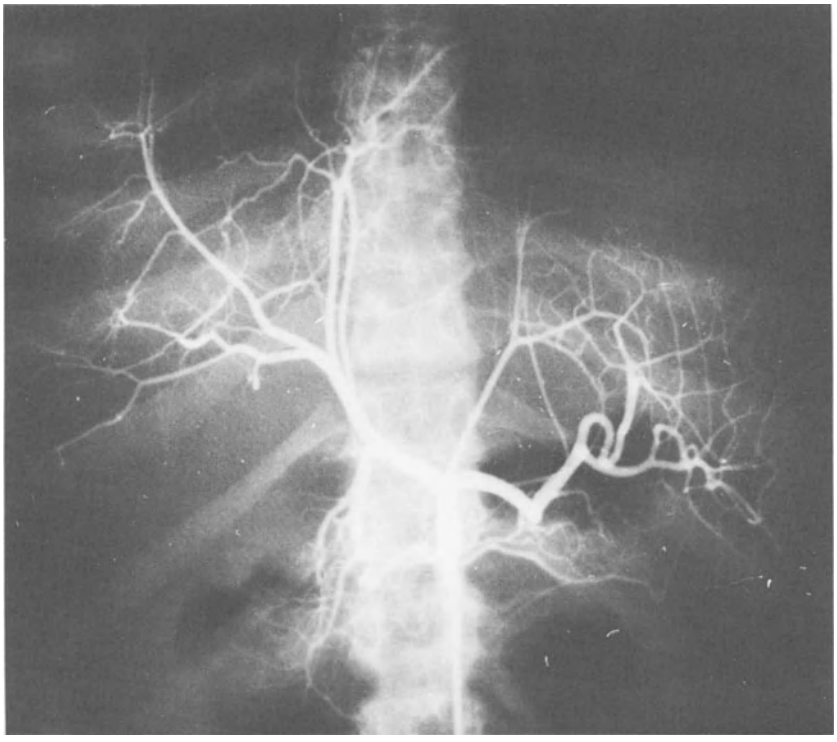
Fig. 9. Atropho-hypertrophic alcoholic cirrhosis in a 51-year-old female referred for variceal bleeding (25–5–79).
9a: Atropho-hypertrophic liver with markedly dilated hepatic artery.
Minimal splenomegaly (15 cm) with very thin and faintly opacified intrasplenic arterial branches.

9b: Dense arterial hepatography.
Very poor splenography with the splenic vein barely visualized.



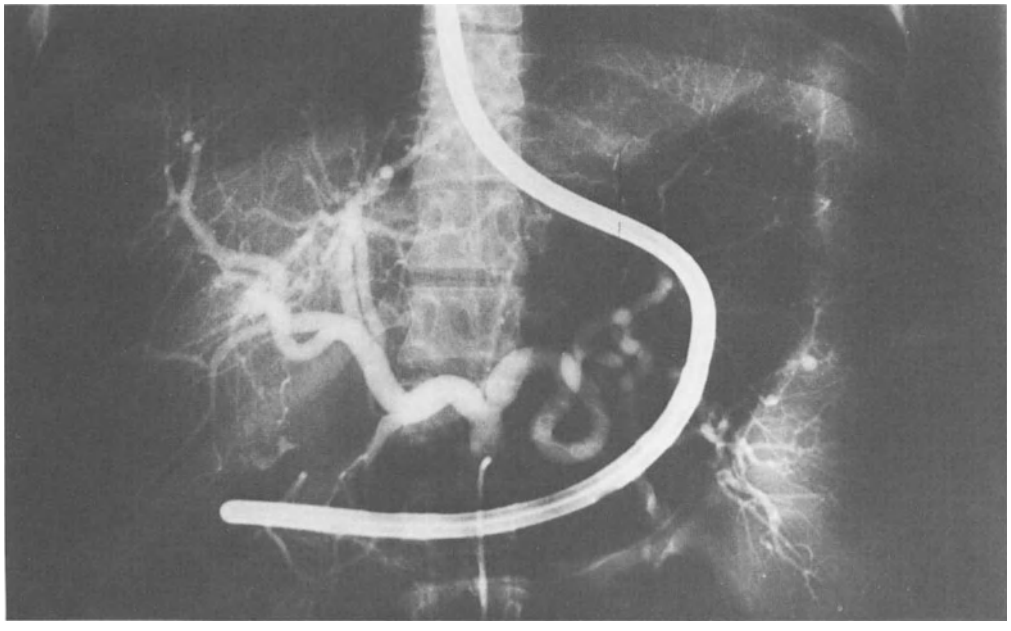
9c

9c: Poor opacification of narrowed intrahepatic portal branches with retrograde flow in the umbilical vein.
Angiographic pattern of a markedly decreased mesenterico-portal flow with almost completely interrupted splenic blood flow resulting from a rapidly progressive intrahepatic obstruction.
The obstruction is more severe than in Fig. 8 as evidenced also by the greater dilatation of the hepatic artery.



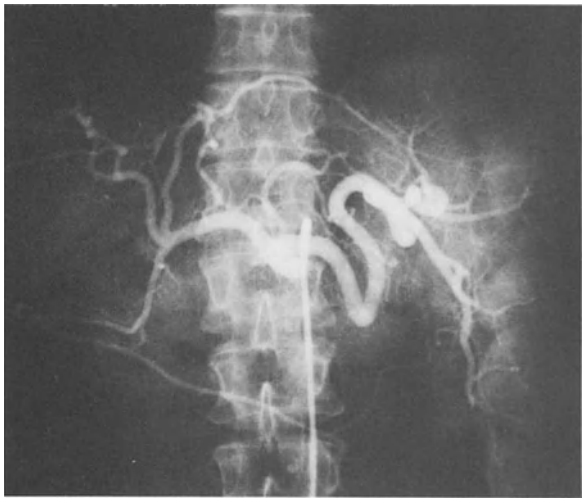
10a

Fig. 10. Budd-Chiari syndrome due to primary hepatic vein thrombosis in a 15-year-old female. Patient referred for unexplained ascitis (1–2–71).
10a: Hypertrophy of the left lobe of the liver with stretched and thinned intrahepatic arteries typical for Budd-Chiari syndrome. Spleen size within normal limits (14 cm) with very thin intrasplenic arterial branches which signifies the slowed splenic arterial flow.

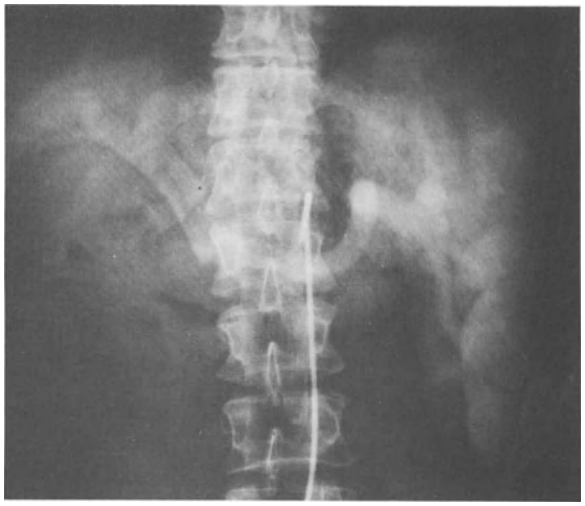


10b

10b: Checking arteriography (30–1–76) 5 years after a portocaval anastomosis.
Owing to the surgical shunt which was shown to be patent, a pronounced splenomegaly (25 cm) with intrasplenic small aneurysms developed as a result of the lowered portal hypertension whereas hepatic artery dilated.
Finally, surgery permitted the markedly reduced hepatic and splenic arterial flow to increase.



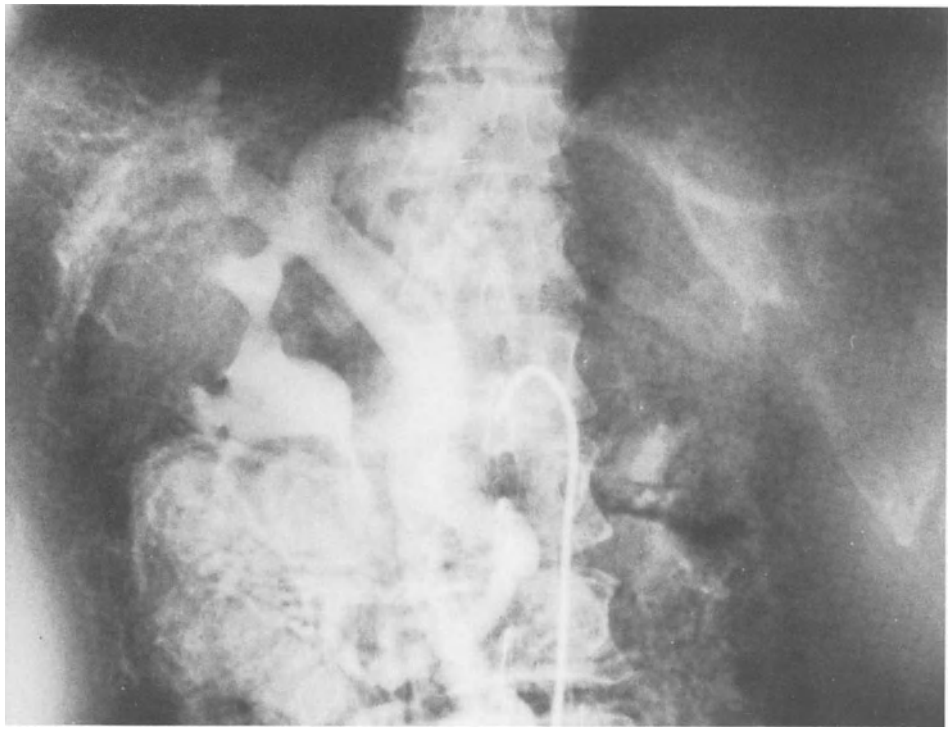
11a



11b

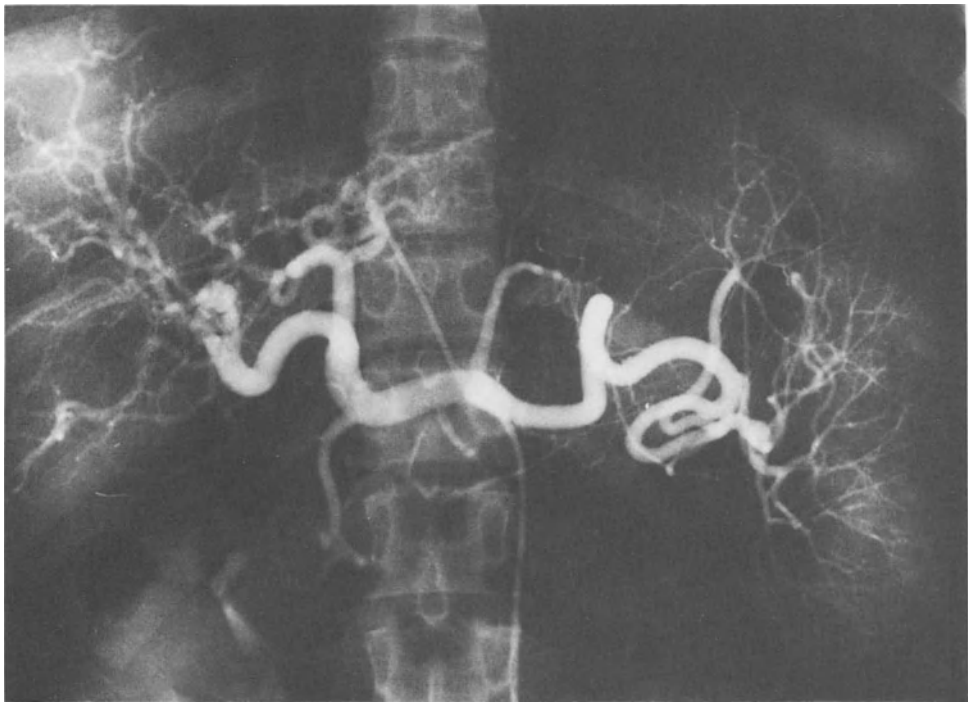
Fig. 11. Post hepatitis-cirrhosis in a 51-year-old male referred for hypersplenism (15–9–81). Neither known previous history of virus hepatitis nor gastro-intestinal bleeding. Only minimal liver function test abnormalities – Hbs Ag + – Laparoscopy: Atrophy-hypertrophic liver with multiple modules.
11a: Atrophic right lobe and hypertrophic left lobe of the liver. Hepatic artery caliber within the normal limits. Markedly enlarged spleen (20 cm) with widened and lengthened splenic artery.

11b: Splenic vein well opacified.

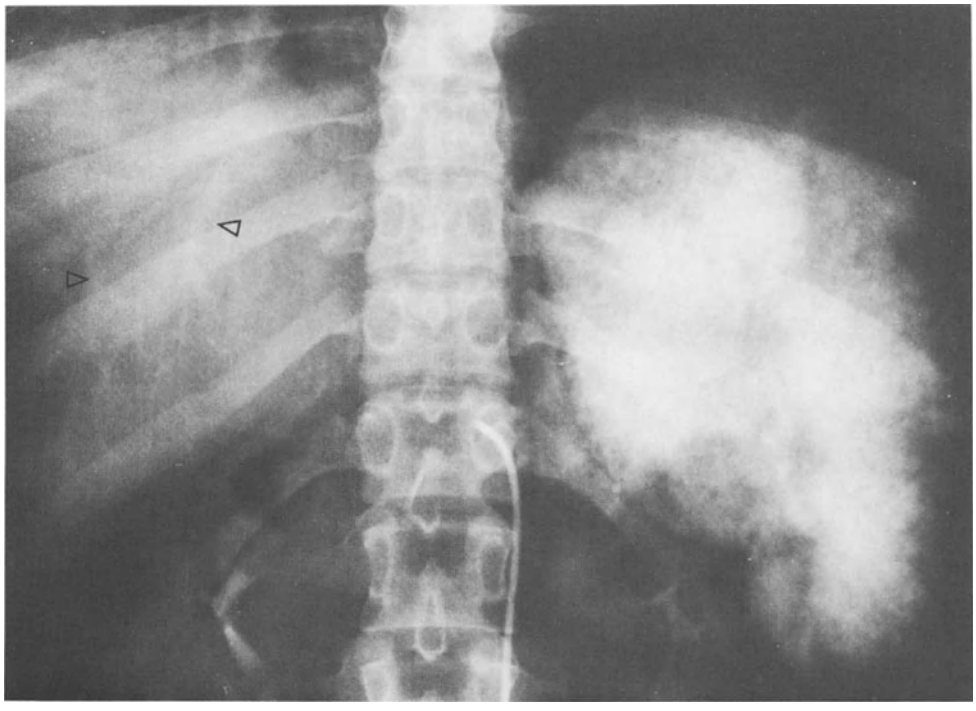


11c

11c: Venous phase of S.M.A. accounting for a relatively good hepatopetal portal flow without hepatofugal flow.

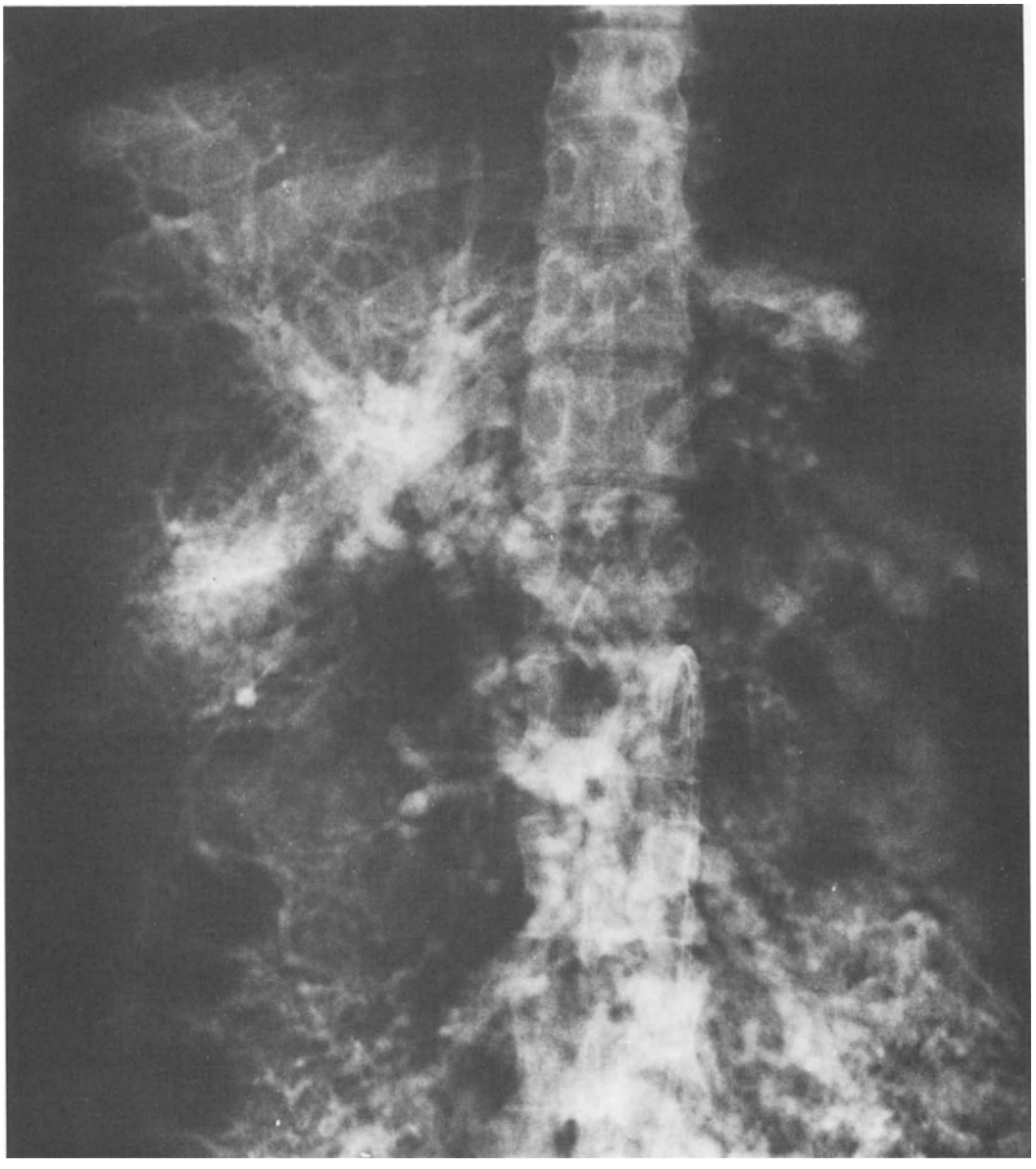


12a



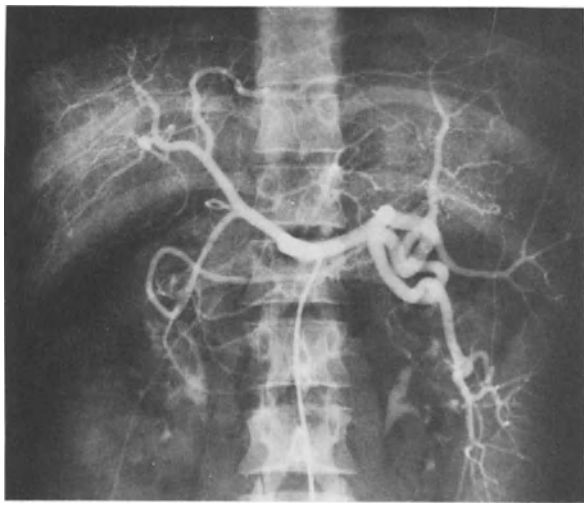
12b

Fig. 12. Portal vein obstruction in a 22-year-old female referred for variceal hemorrhage (9–10–75) and probably resulting from pylephlebitis during pregnancy 2 years earlier.
12a and b: Pronounced splenomegaly (20 cm) with relatively thin intrasplenic arterial branches accounting for a slowed splenic blood flow rate as demonstrated by the prolonged splenography without opacification of the venous outflow.
Markedly dilated hepatic artery accounting for a greatly reduced hepatopetal portal flow. The faint opacification of the hepatic veins (12b – ↑) permits the prehepatic site of the obstruction to be anticipated.

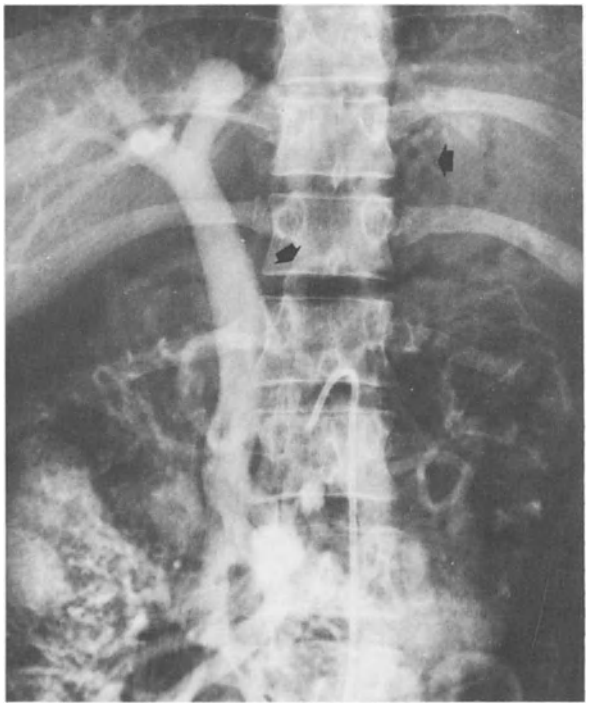


12c

12c: Complete obstruction of the mesenterico-portal axis with predominant hepatopetal portal flow associated with cavernous transformation of the portal vein.



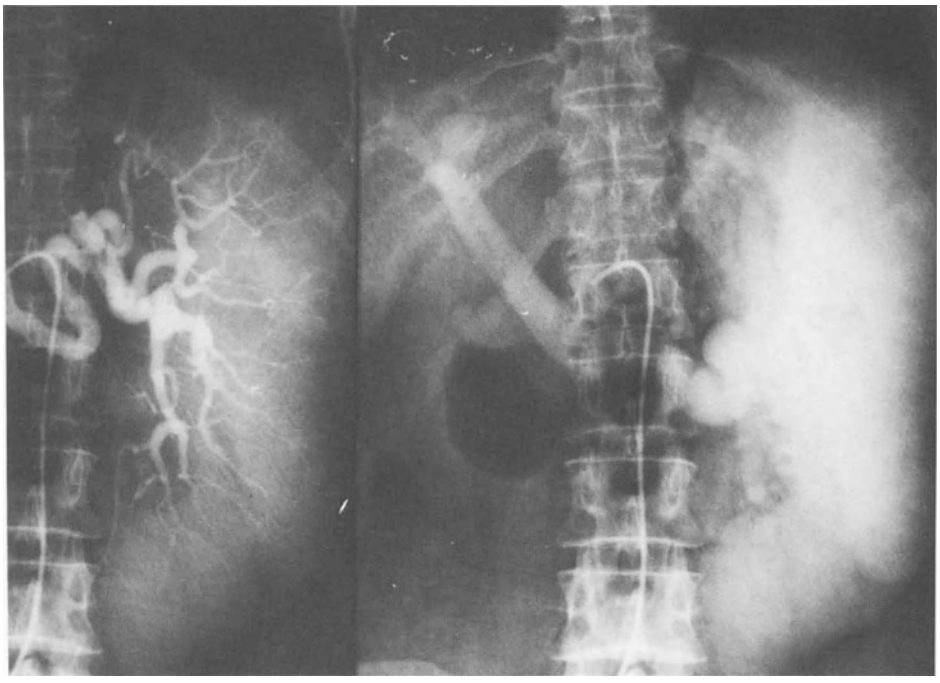
13a



13b

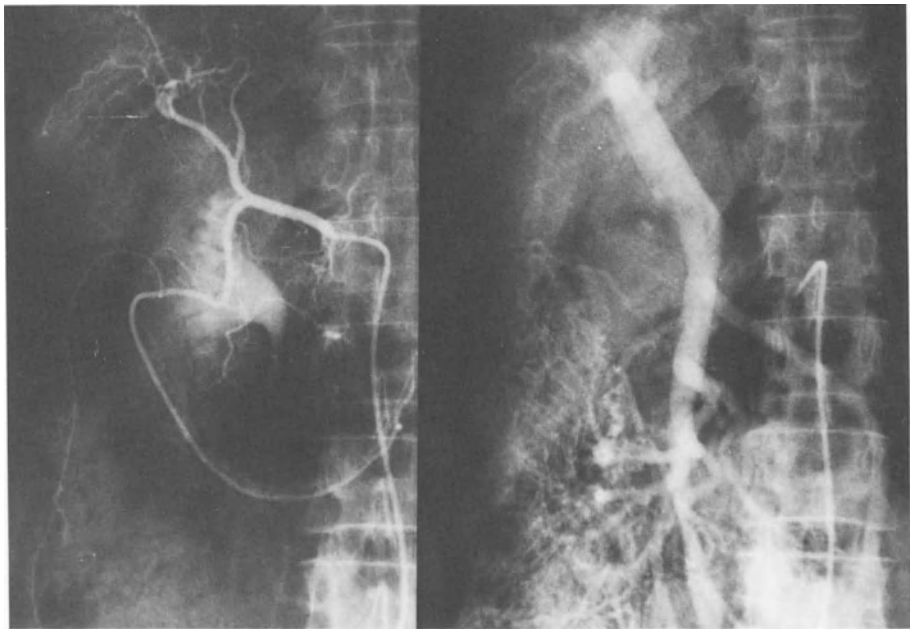
Fig. 13. Wilson's disease with moderate, long-standing and slowly progressive portal hypertension in a 23-year-old male. Patient referred for esophageal varices bleeding (24–5–74). Corrected sinusoïdal pressure: 12 cm saline.
13a: Huge splenomegaly (30 cm) with widened and lengthened splenic artery and a small intrasplenic aneurysm (⌓)
Moderate dilatation of the hepatic artery and its intrahepatic branches.

13b: Venous phase of S.M.A.
Secondary and tertiary intrahepatic portal branches faintly visualized, which signifies the moderately reduced portal inflow. Very faint opacification of left gastric vein and varices (↑).



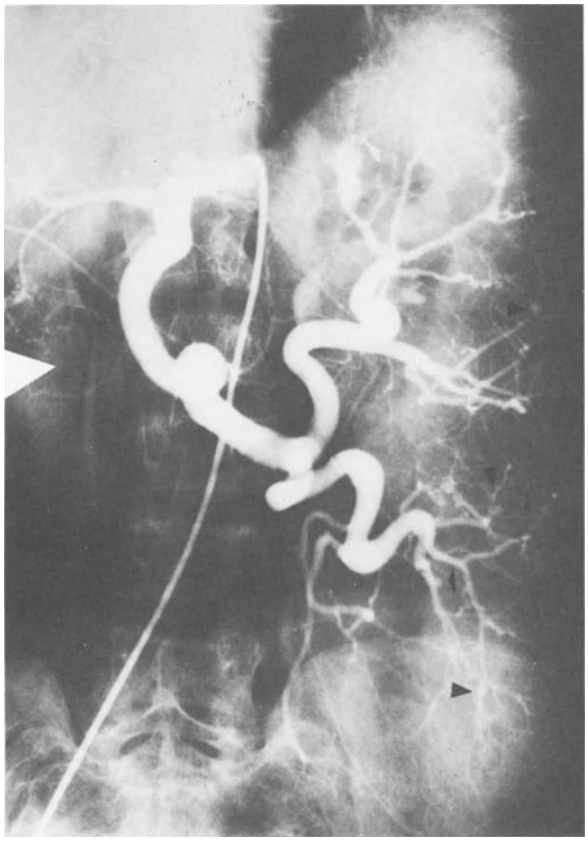
14a

Fig. 14. Slowly progressive alcoholic cirrhosis in a 49-year-old female (9–5–83) known to have experienced several episodes of variceal bleeding, ascitis and jaundice for 12 years. Hypersplenism. Child's score: B7.
14a: Huge splenomegaly (26 cm) with dilated and lengthened splenic artery-dense splenography and portal vein well visualized.

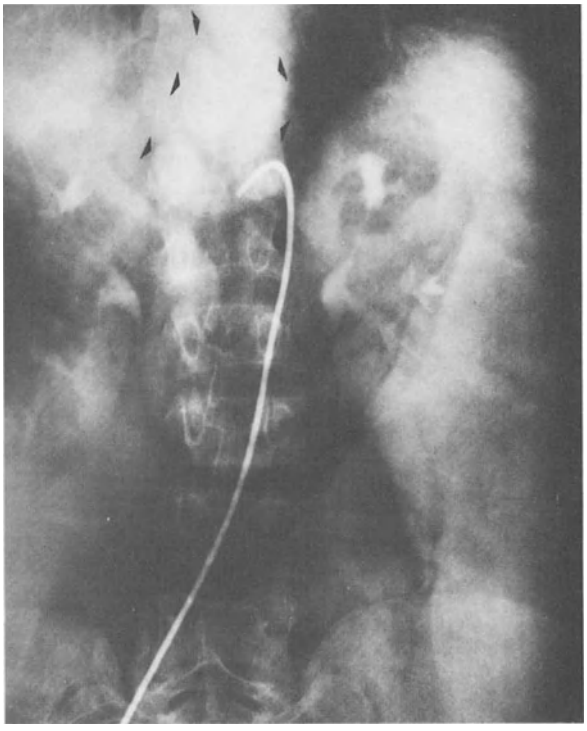


14b

14b: Atrophic liver with normally thin intrahepatic branches. Good opacification of the portal vein with only minimal retrograde flow in the inferior mesenteric vein which signifies a predominant hepatopetal portal flow.
Typical angiographic pattern for a moderate and slowly progressive intrahepatic obstruction.
Splenectomy and proximal spleno-renal anastomosis performed on 23–5–83:
* Portal pressure: 16 cm saline
* Fibro congestive spleen weighing 1,000 g:
* Liver biopsy: annular fibrosis with neither inflammatory reaction nor liver cell lesion:
Pathological pattern of moderate and stationary cirrhosis.



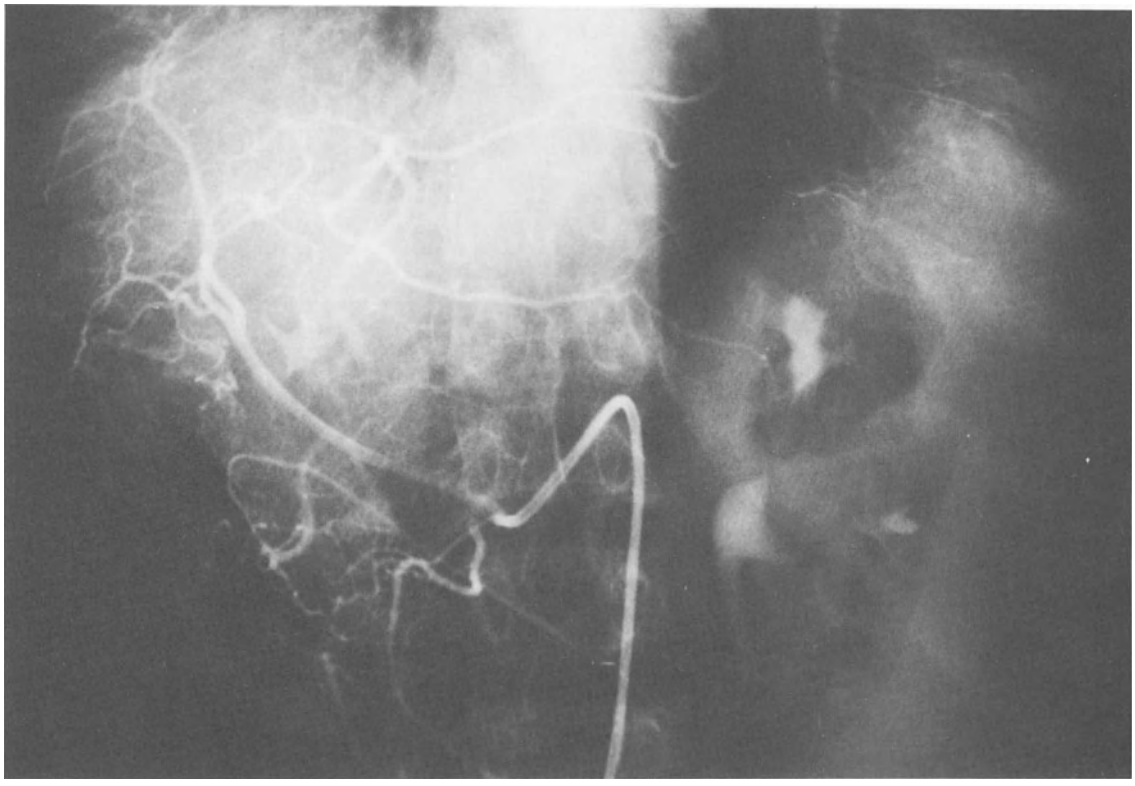
15a



15b

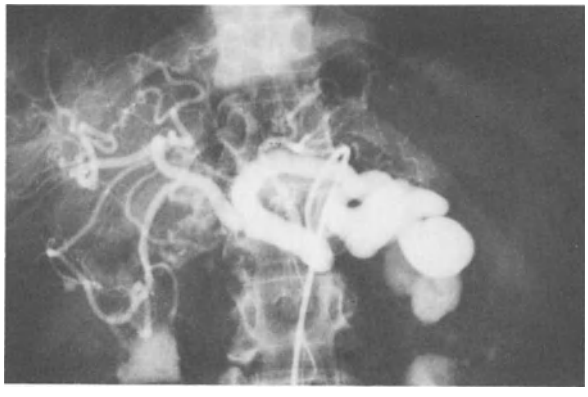
Fig. 15. Splenomegaly associated with hypersplenism in a 39-year-old female originating from North Africa (6–8–75).
15a: Huge splenomegaly (27 cm). Markedly dilated splenic artery with somewhat irregular caliber and multiple small aneurysms of the intrasplenic arterial branches (↑).

15b: Faint opacification of the markedly dilated spleno-portal axis together with very large esophageal varices (↑).



15c

15c: Selective right hepatic artery opacification: atrophic right lobe and opacification of a markedly hypertrophic left lobe through anastomosis between both hepatic arteries.
Splenectomy (29–8–75):
Portal pressure: 20 cm saline. Spleen weight: 920 g. Pathology:
– Moderate portal fibrosis and diffuse inflammatory reaction in the liver;
– Fibro-congestive spleen;
– Splenic arterial wall pattern suggesting fibro-muscular dysplasia.



16a

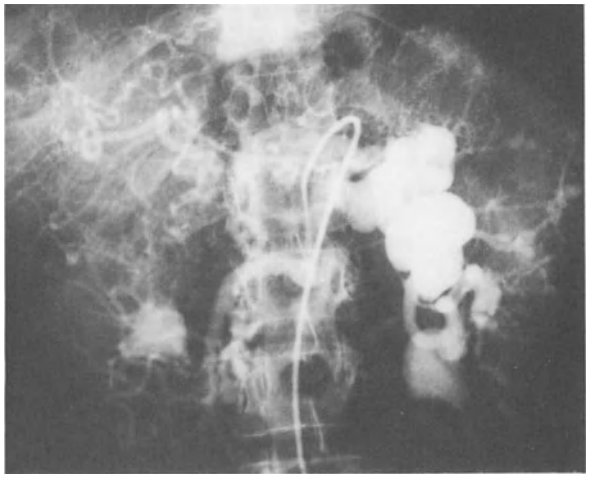
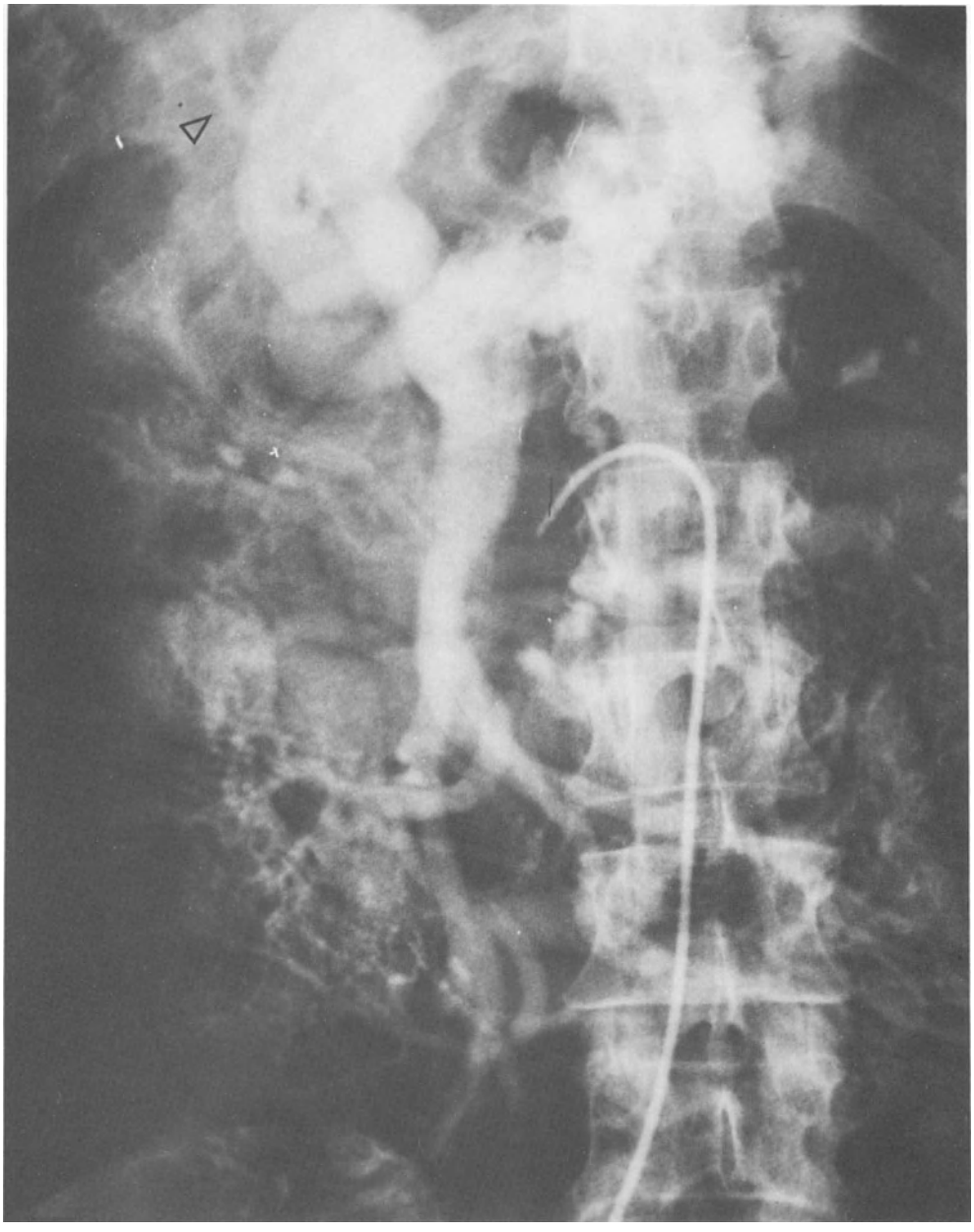
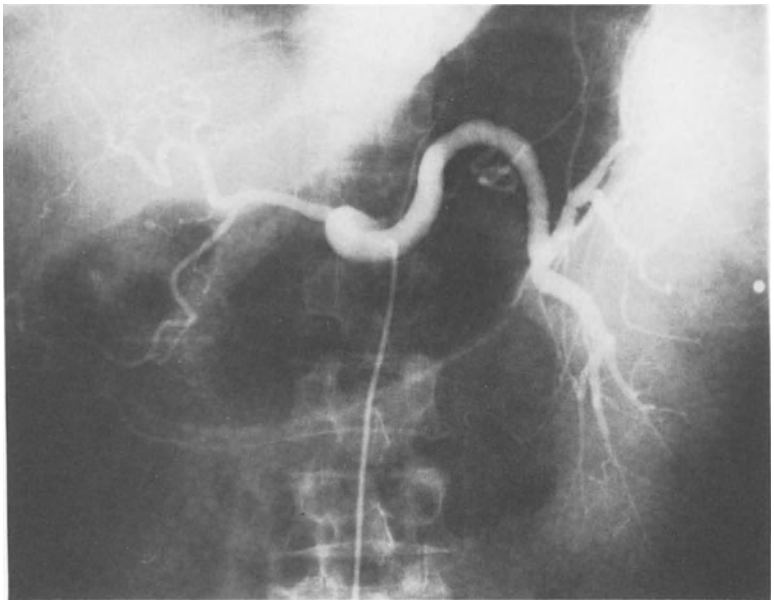


Fig. 16. Atrophic cirrhosis of unknown origin with hypersplenism and hepatic failure in a 57-year-old female.
16a: Marked atrophy of the liver with moderate dilatation of the hepatic artery.
Pronounced splenomegaly (19 cm) with large aneurysms of the splenic artery and several smaller ones within the splenic parenchyma.



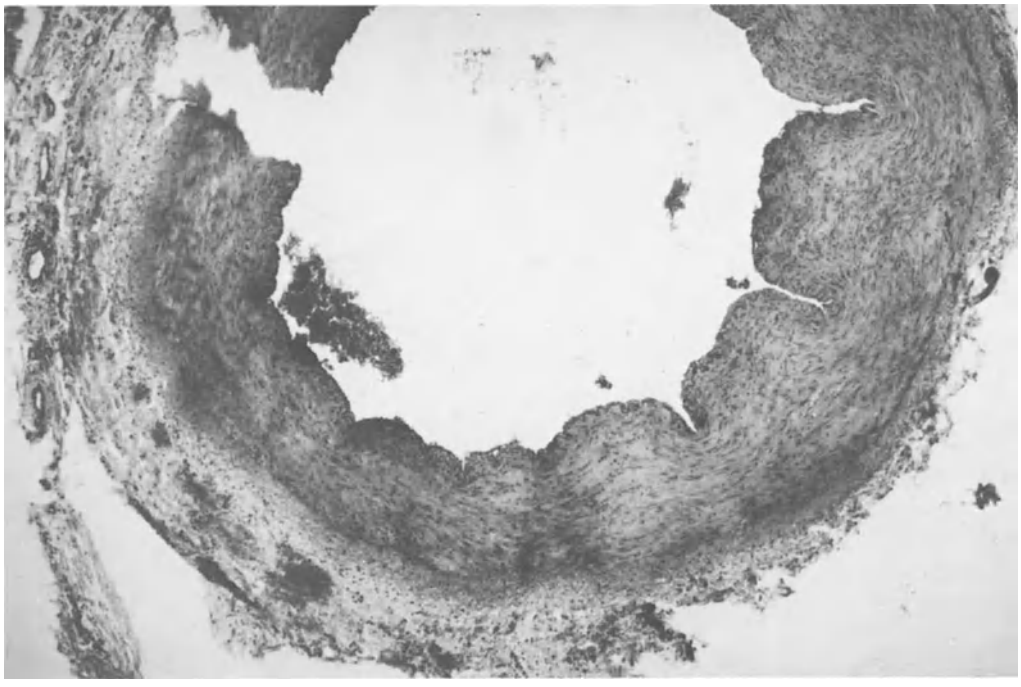
16b

16b: The narrowed right portal vein (↑) signifies the low hepatopetal portal flow whereas a predominant hepatofugal flow through a dilated umbilical vein and gastro-esophageal varices is evidenced.
– Splenectomy (6–11–79)
– Portal pressure: 31 cm saline
– Spleen weight: 475 g
– Atrophic and micro modular cirrhosis confirmed by liver biopsy.



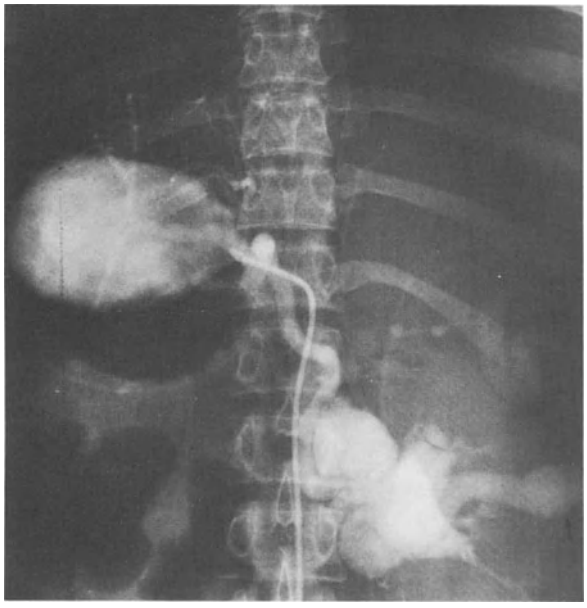
17a

Fig 17. Hepatic fibrosis of unknown origin in a 61-year-old male referred for esophageal varices bleeding (6–1–68).
– Portal pressure measured through splenoportography 25 cm saline
– Splenectomy was performed (23–1–68) and the patient died 4 years later from hepatic failure
– Liver biopsy: mild inflammatory peri-portal fibrosis.
17a: – Huge splenomegaly (29 cm)
– irregularly dilated splenic artery with the 'string of bead' pattern suggesting fibro-muscular dysplasia
– Normal angiographic liver pattern.

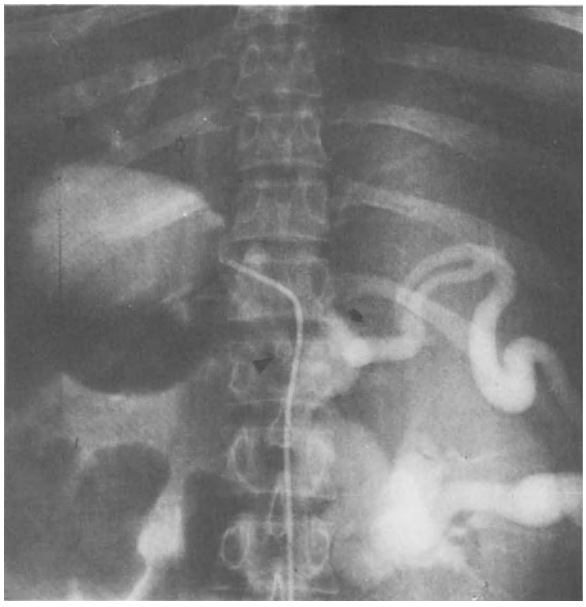


17b

17b: Pathological examination of the splenic artery (H.E.S. × 25 – Pr. M. Houcke)
– Irregular thickening and fibrosis replacement of the medial layer with invagination of the internal elastic lamella. Pathological pattern typical for arterial fibro-muscular dysplasia of the medial type.



18a

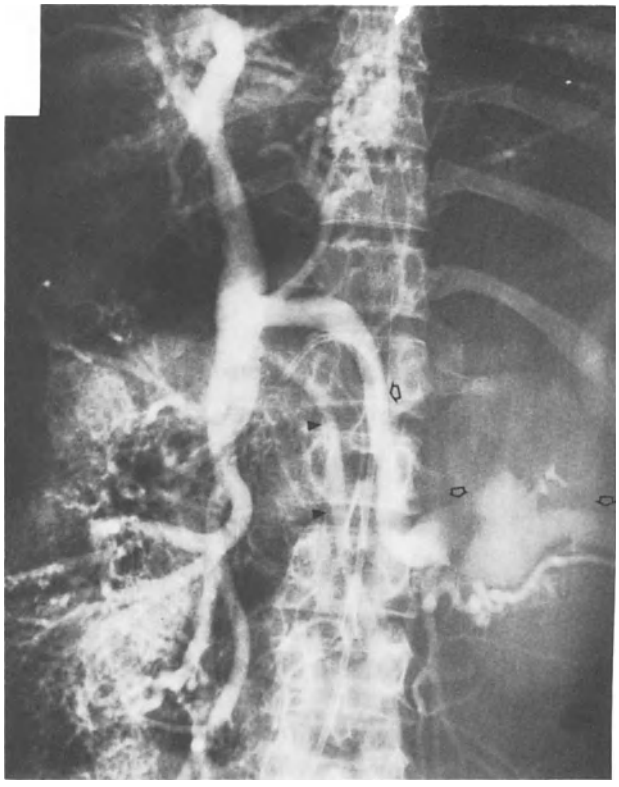


18b

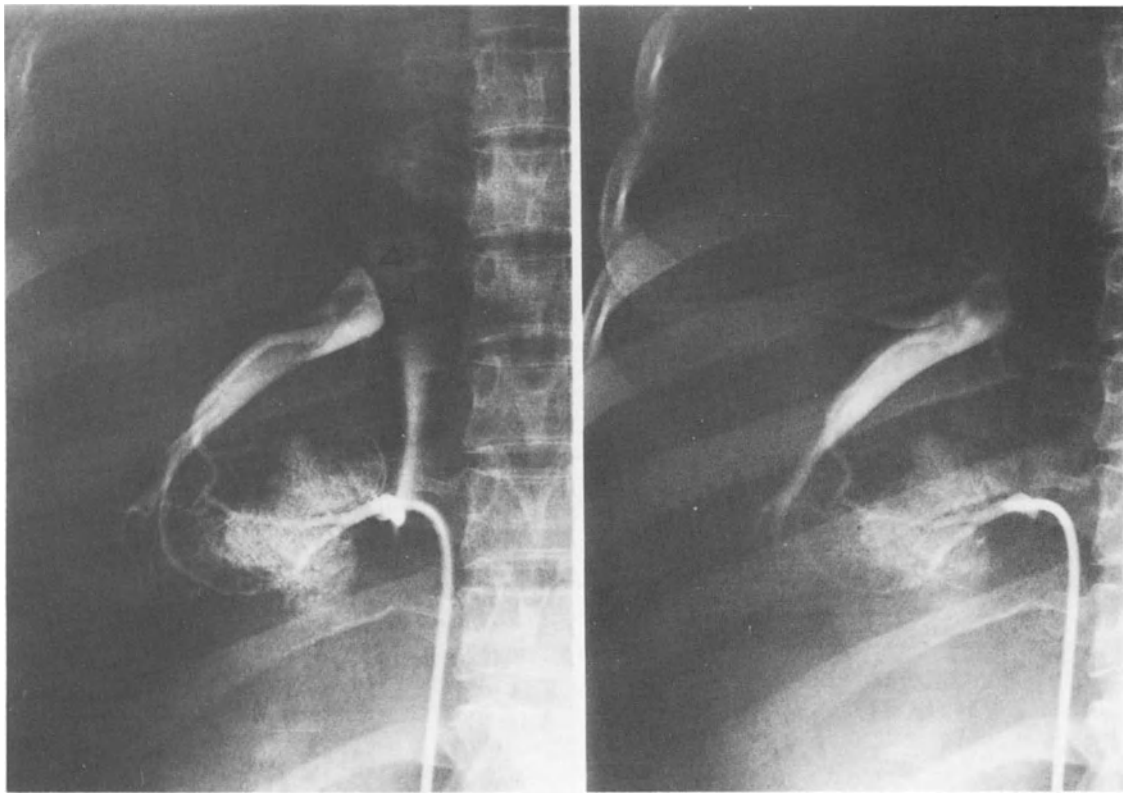
Fig. 18. Congenital Budd-Chiari syndrome in a 32-year-old female originating from West Indees. This patient who never experienced any trouble before was found to have splenomegaly with hypersplenism following pregnancy (8–5–74).
18a: Coeliac arteriography: huge splenomegaly occupying the whole left half of the abdomen and atrophic liver.
Several very large splenic artery aneurysms: one of them proximally located is seen in front of hepatic artery (⌓) whereas others are located in the splenic hilum and communicate with the splenic vein (↑).

18b: A few seconds later, the splenic vein opacification resulting from this aneurysmal arterio-venous fistula is seen to fill a spontaneous spleno-renal anastomosis (↑) and inferior vena cava (⌓) as well as the splenoportal axis in a hepatopetal direction (▲).

18c

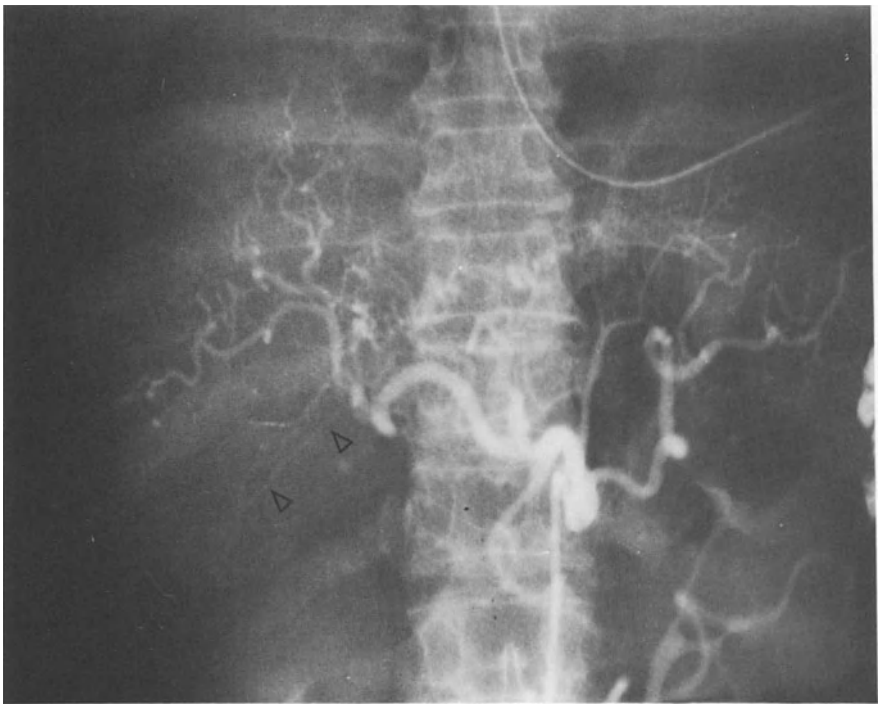


18c: Venous phase of S.M.A. shows a patent portal vein with a predominant hepatofugal flow through left gastric vein, inferior mesenteric vein (▲), splenic vein and splenorenal anastomosis (⌓).



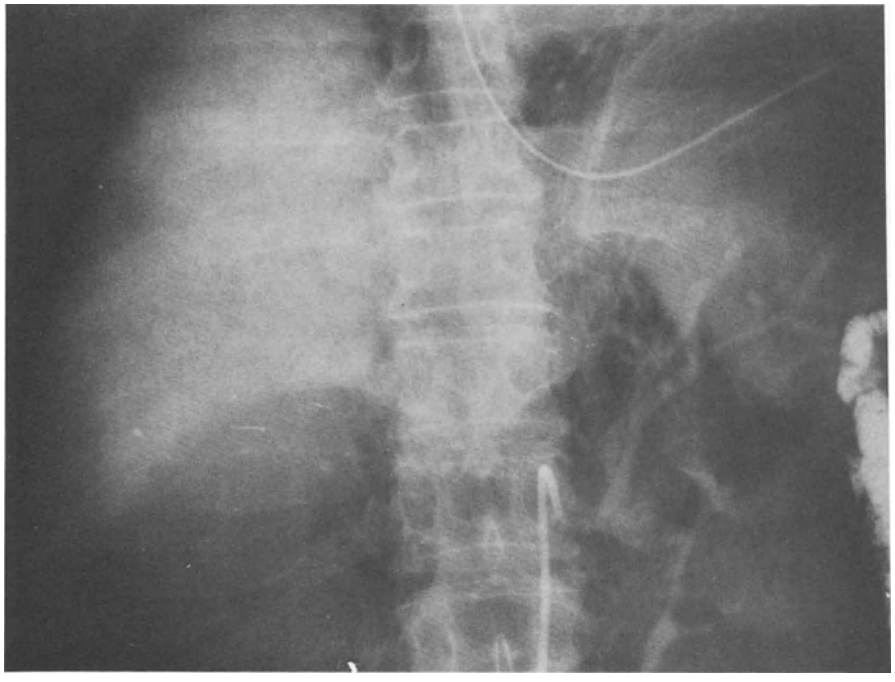
18d

18d: Hepatic venography: only a small accessory hepatic vein can be catheterized, allowing the main right hepatic vein to be opacified through intrahepatic anastomosis.
The distal end of the right hepatic vein does not open into the inferior vena cava from which it remains separated by a thin lucent stripe (△), suggesting a membranous or web-like obstruction. As a result the main right hepatic vein remains opacified for a few seconds after the end of the injection.
Splenectomy: fibro-congestive spleen weighing 2,000 g.
Liver biopsy: periportal sclerosis with acute congestive liver and dilated sinusoids.



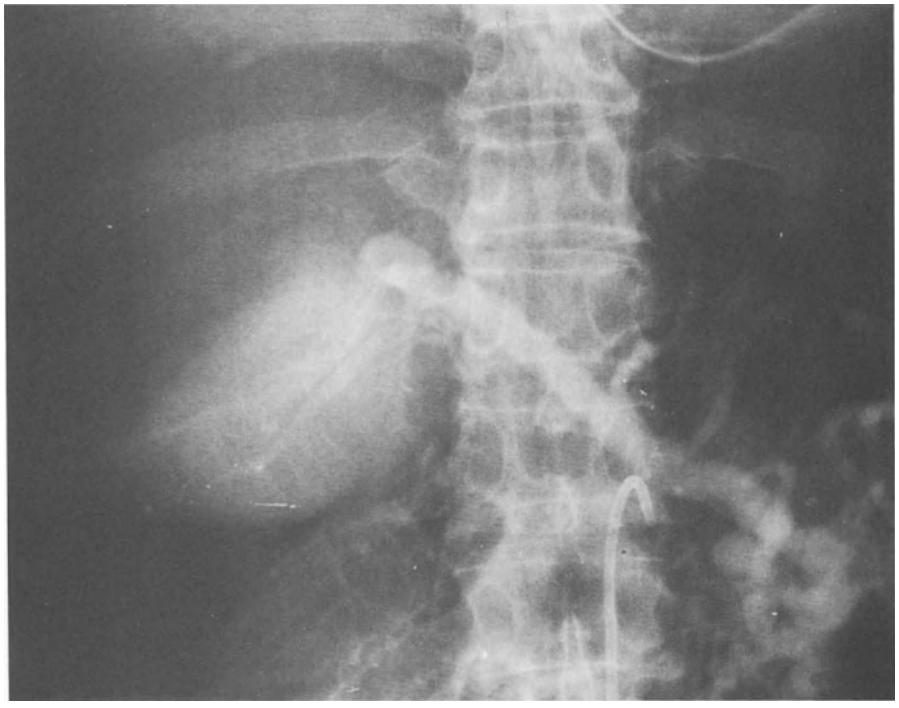
19a

Fig. 19. Severe acute pancreatitis with ascitis, severe involvement of the transverse colon and portal vein thrombosis in a 65-year-old male (25–8–77).
19a: Moderate dilatation of the segmental intrahepatic arterial branches except the artery of the segment 5 which is very thin (↑). (All branches of the coeliac artery are probably narrower than normally because of acute pancreatitis and hypotension.)



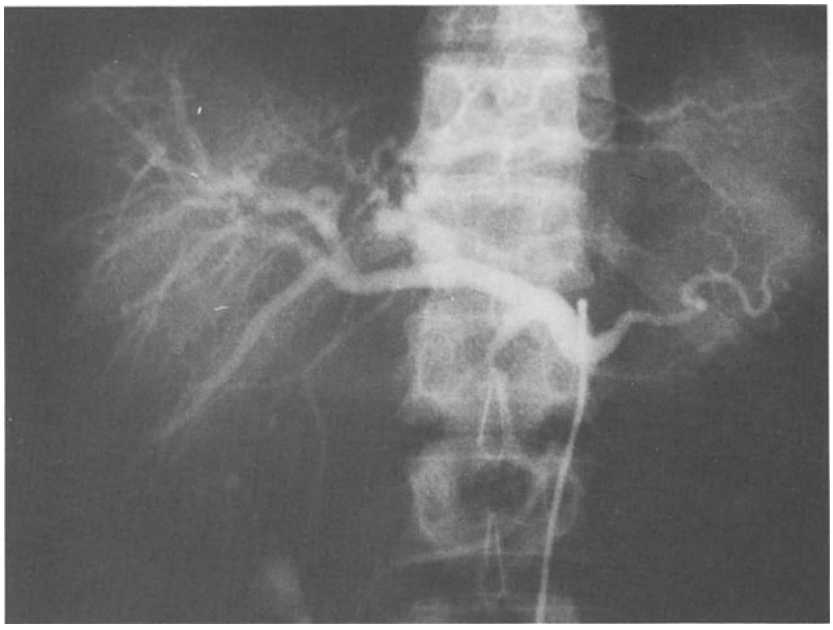
19b

19b: Venous phase of coeliac arteriography shows the arterial hepatography to be abnormally dense in all hepatic territories where the arterial branches are dilated, whereas a normally faint hepatography is seen in the segment 5.



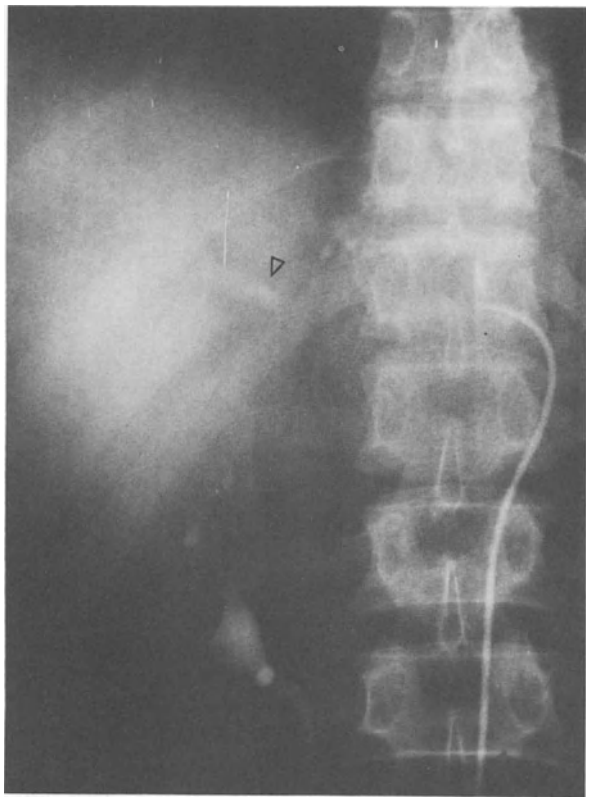
19c

19c: Venous phase of S.M.A. demonstrates a patent portal stem with irregular walls due to incipiens thrombosis. Portal branches of the segment 5 remain patent resulting in a normally dense portal hepatography in this territory, whereas all other intrahepatic branches are completely obstructed.
Arterial and portal hepatography result in a 'mirror image' demonstrating that arterial flow increases to compensate for the lost portal flow.



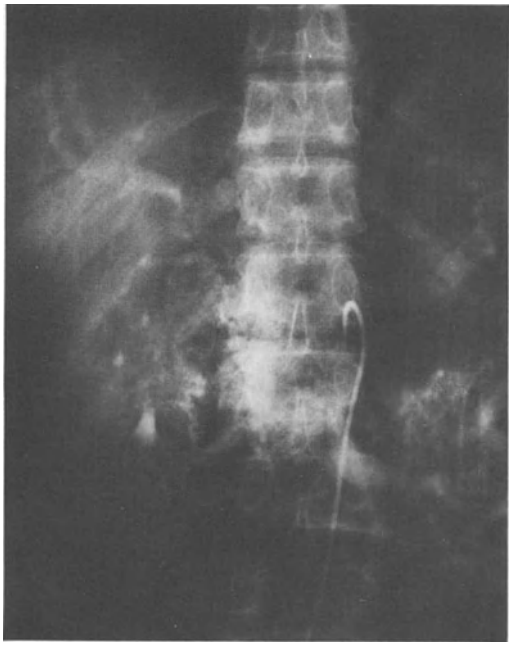
20a

Fig. 20. Chronic pancreatitis previously treated by spleno-pancreatectomy in a 41-year-old male.
Patient referred for a recurrent bout of pancreatitis (20–3–79).
20a. Markedly dilated hepatic artery which signifies the reduced portal inflow.



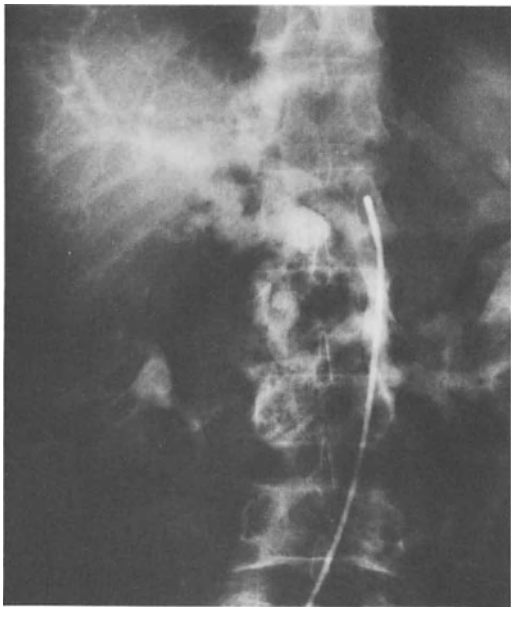
20b: 5 sec later retrograde filling of the portal vein through intrahepatic arterioportal communications is clearly documented (↑).

20b



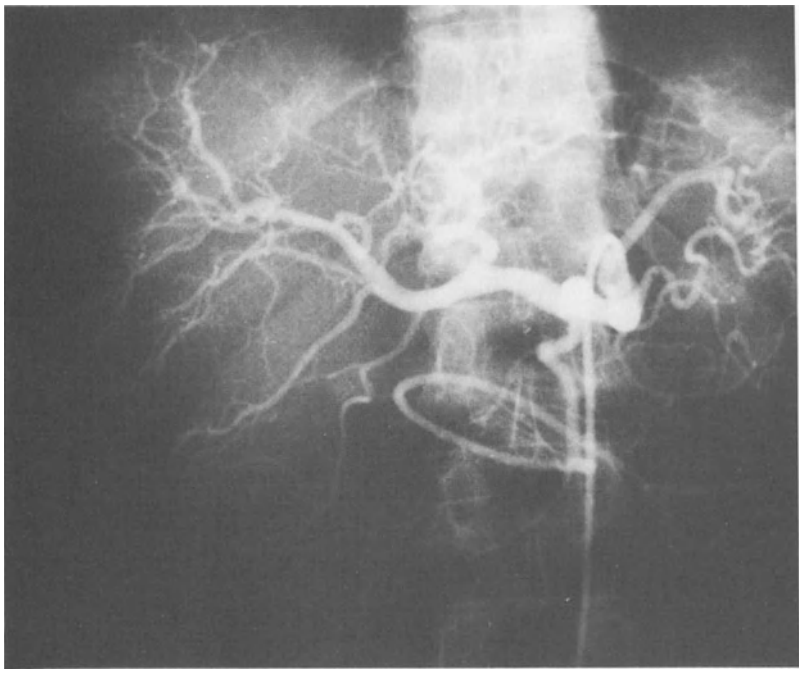
20c

20c: Venous phase of S.M.A. shows thrombosis of the mesenterico-portal axis with little hepatopetal collateral flow resulting in only faint opacification of the intrahepatic portal veins.
Two necrotic and infected pancreatic collections are treated by surgical external drainage (29–3–79).



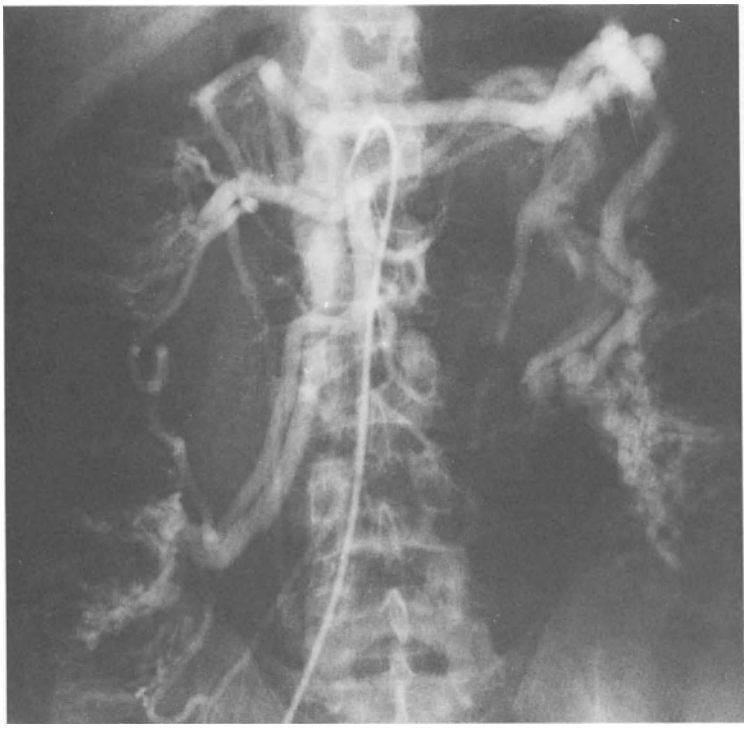
20d

20d: Checking arteriography performed 6 months later (14–9–79) shows on the venous phase of S.M.A. that the mesenterico-portal axis is no longer obstructed. Not only is it partially patent but a wide hepatopetal collateral way occurred through the pancreatico-duodenal veins. Owing to this a good filling of the intrahepatic portal venous bed is visualized.



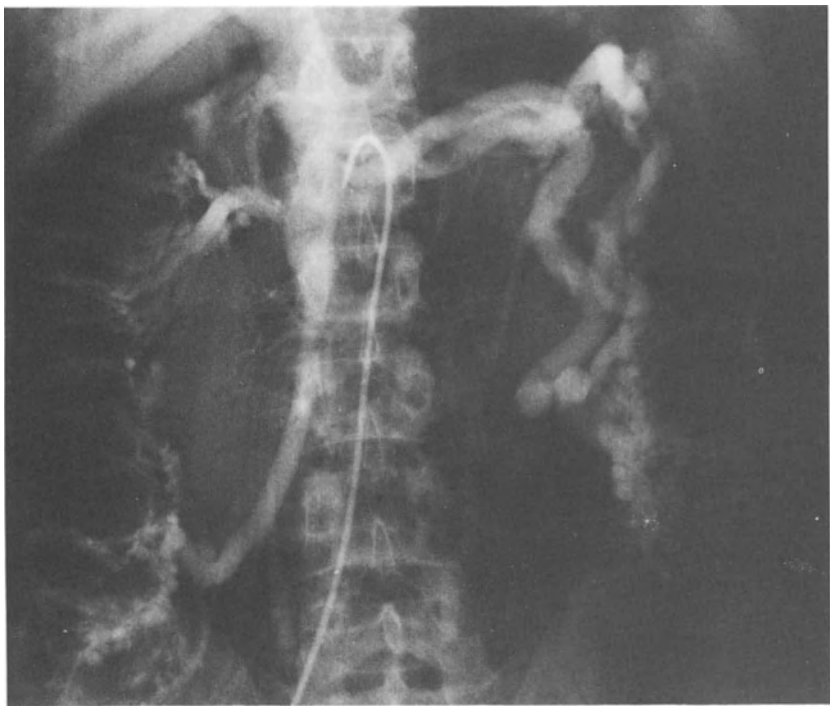
20e

20e: Coeliac arteriography (14–9–79) shows the hepatic artery to be less dilated than previously, mainly when comparing the diameter of intrahepatic arterial branches, which signifies the better portal inflow.



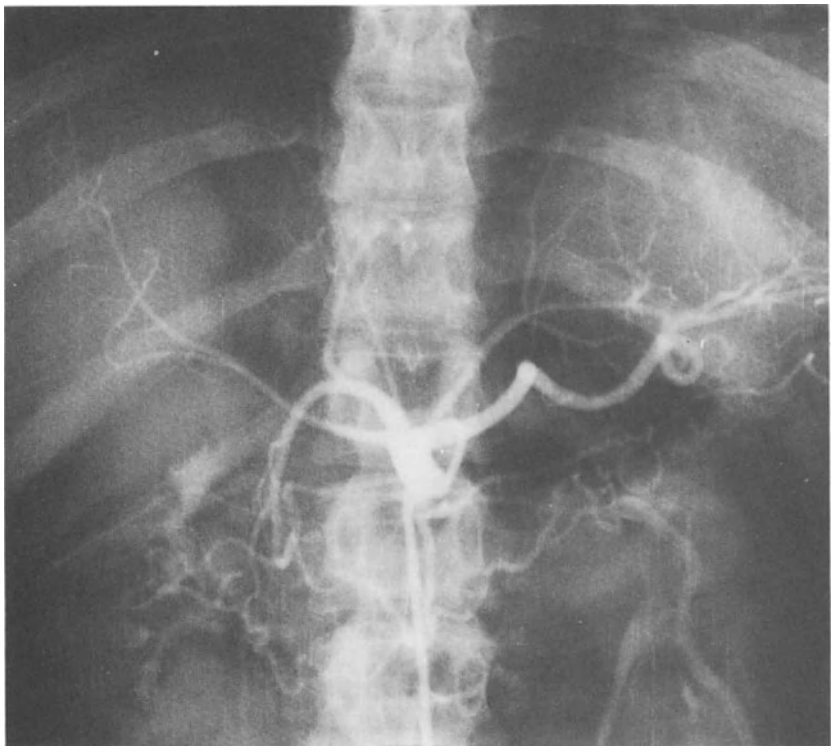
21a

Fig. 21. Diffuse and severe angiodysplasia of the colon in a 29-year-old male. Patient referred for anemia and intestinal bleeding (9–11–82).
21a: S.M.A. shows markedly widened right and left colic arterial branches with immediate filling of the draining veins through wide and numerous arterio-venous fistulae.



21b

21b: 2 seconds later the portal vein is densely opacified.



21c

21c: On coeliac arteriography hepatic artery is abnormally thin and its peripheral intrahepatic branches are not opacified, which suggest a decreased hepatic arterial flow.
A two-fold increase of the cardiac output could be measured. Normal liver and normal portal pressure at surgery.

4. PRESINUSOIDAL OBSTRUCTIONS

I. Proper angiographic signs

Prehepatic obstructions are the most common form of presinusoidal portal hypertension, usually resulting from portal vein or splenic vein thrombosis. Unlike postsinusoidal obstructions, a normally low pressure portal venous bed is preserved beyond the obstacle, which is responsible for some specific hemodynamic changes.

1. Arterial changes

The portal vein obstruction is known to determine the more pronounced hepatic arterial dilatation as a result of decreased portal flow (Fig. 12). Moreover, it is probably a favorable condition for the intrahepatic arterio-portal shunting to develop as a pressure drop occurs in the portal veins located beyond the obstruction. Indeed portal vein opacification through hepatic artery injection of contrast medium has been documented in some cases of portal thrombosis (13).

It is, however, an uncommon finding in clinical circumstances as sinusoidal pressure and blood flow rate within the liver are normal. In fact we found out this hemodynamic change only as a transient phenomenon at an early stage of a portal thrombosis (Fig. 20).

In most cases, the contrast medium injected into the hepatic artery normally flows through the sinusoids to join the hepatic veins. It is the reason why the retrograde filling of the portal venous bed located beyond the obstruction is an uncommon finding of hepatic arteriography in pre-hepatic obstruction.

On the contrary, the hepatic vein opacification is not such an uncommon finding after contrast medium injection into the hepatic artery in these cases. Indeed, the higher the arterial flow, the more the sinusoids filled with a more concentrated contrast medium. In addition, the lower the portal flow, the less diluted the contrast medium injected through the arterial route.

As a result of this the dense arterial hepatography is not weakened by the unopacified portal blood, and because of the normal blood flow rate within the liver, a good opacification of the hepatic veins may be seen (Figs. 12, 22, 23). On the contrary, in case of post-sinusoidal obstruction, the increased intrahepatic resistances result in blood stasis and dilution of the contrastmedium so that the hepatic veins cannot be visualized even though the arterial flow is increased. Another indirect sign of prehepatic obstructions is the normal morphologic appearance of the liver with a normal distribution of the intrahepatic branches which can be well documented by hepatic arteriography (Figs. 12, 22, 23, 24). In conclusion, a normal morphologic pattern of the liver associated with a usually pronounced hepatic arterial dilation and occasionally hepatic vein opacification is the typical sign of a prehepatic obstruction on hepatic arteriography.

It must be stressed that the diagnosis of prehepatic obstruction may be suggested by C.T. scan with bolus injection of contrast medium. Owing to the usually pronounced dilatation of the hepatic artery, the intrahepatic arterial branches are often widened enough to be seen on the first slice corresponding to the arterial phase, whereas they are too thin to be visualized in normal persons. Moreover, the venous collateral circulation together with a thrombus within the portal vein may be also documented 20 seconds later (Fig. 23).

2. Venous changes

The hepatopetal porto-portal collateral venous circulation is typical of presinusoidal obstructions (Figs. 12, 23, 24, 25). In most cases, the hepatopetal collateral flow is predominant as compared to the hepatofugal circulation so that portal hypertension

is less severe than in post-sinusoidal obstructions, and bleeding from esophageal varices is less likely to occur. The hepatopetal collateral pathways depend on the site of the obstruction.

Splenic vein obstruction. An occluded splenic vein is easily bypassed owing to the left gastric veins running along the lesser curvature of the stomach as well as the veins of the greater curvature (gastro-epiploîc veins) so that the splenic venous outflow may remain normal or near normal (Fig. 26). The splenic venous stasis mainly depends on the length of the obstruction so that involvement of the confluence of the splenic, mesenteric and portal veins always results in a decreased splenic venous outflow.

Mesenteric vein obstruction. When the obstacle is located in the superior mesenteric vein (in case of an enlarged processus uncinatus of the pancreas for instance) bridging collaterals are usually formed over the veins running along the intestine to either the right superior colic vein or the first jejunal vein which, in some cases, drains into the superior mesenteric vein juste before it joins the spleno-portal axis (Fig. 27).

Obstruction of the spleno-mesenterico-portal confluence. In this case the superior mesenteric venous blood drains towards the portal vein through the dilated pancreatico-duodenal veins (Fig. 28).

Portal vein obstruction. Portal vein obstruction produces an extensive network of venous channels in the hepato-duodenal ligament, usually described as the cavernous transformation of the portal vein. It allows the splenic and mesenteric venous blood to reach the intrahepatic portal veins through the subcapsular portal venous network (Fig. 29). However, in spite of this porto-portal collateral network, the portal flow is markedly decreased and hepatofugal porto-systemic collaterals occur as in case of post-sinusoidal obstruction.

It should be noted, however, that a portal vein thrombosis associated with a post-sinusoidal obstruction, such as cirrhosis, cannot be misinterpreted as a primary portal vein thrombosis because of the lack of hepatopetal collateral circulation (Fig. 25).

II. The main cause of prehepatic obstructions

1. Pancreatic diseases

Pancreatitis and pancreatic carcinoma are the most common causes responsible for compression or thrombosis of the prehepatic portal system (39).

On the basis of 400 cases of pancreatitis studied by arterioportography, the splenic, portal or mesenteric venous involvement was found to be as high as 55%. However, this venous narrowing or occlusion produces only a benign portal hypertension in most cases, giving rise to bleeding from esophago-gastric varices in only 4.5% of the patients.

Splenic vein involvement is the most frequent, occurring in 44% of the cases as the result of lesions located in the body and the tail of the pancreas. Obstruction of the spleno-mensenterico-portal confluence occurs in 28% of the cases and is related to a pancreatic isthmus involvement. The superior mesenteric vein may be involved (14%) by an enlarged processus uncinatus of the pancreas while the splenoportal axis remains normal. Portal vein occlusion occurs in 14% of the cases due to pancreatic head involvement (Figs. 19, 20, 22, 23, 26, 27, 28, 29).

2. Portal vein thrombosis

Portal vein thrombosis is less frequent; its etiology often remains uncertain although neonatal omphalitis is a frequently suspected causative condition. In rare cases, acute portal vein thrombosis was reported to give rise to mesenteric infarctus.

3. Other causes

Numerous and various other causes have been reported to involve the prehepaptic portal venous system such as: retroperitoneal tumors, lymph node tuberculosis (40) (Fig. 30), and chronic volvulus of the small bowel (41) (Fig. 31).

III. Intrahepatic presinusoidal obstructions

The main causes of intrahepatic presinusoidal

obstructions are: schistosomiasis, biliary cirrhosis, congenital hepatic fibrosis (Fig. 32), liver tumor in rare cases, and portal fibrois.

Most of these diseases are very uncommon. The most frequent is probably the portal fibrosis, also described as idiopathic portal hypertension, which raises an interesting diagnostic and physiopathologic problem and will be discussed later.

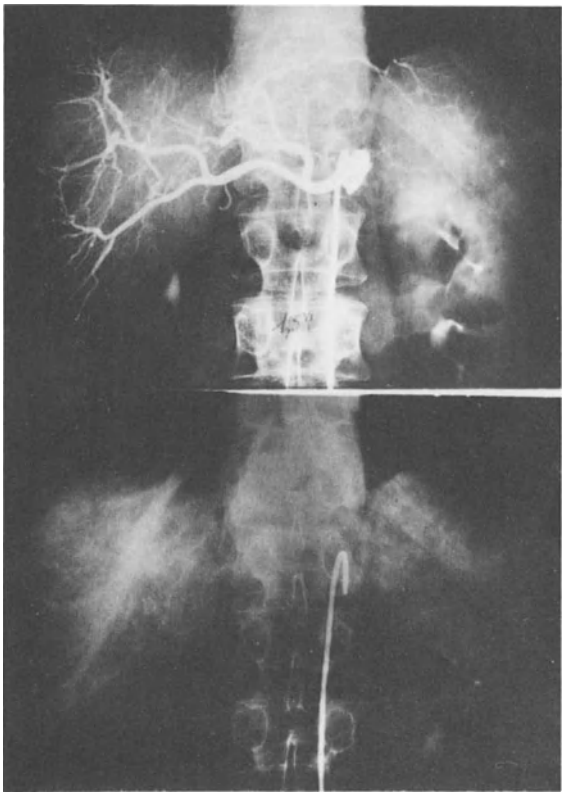
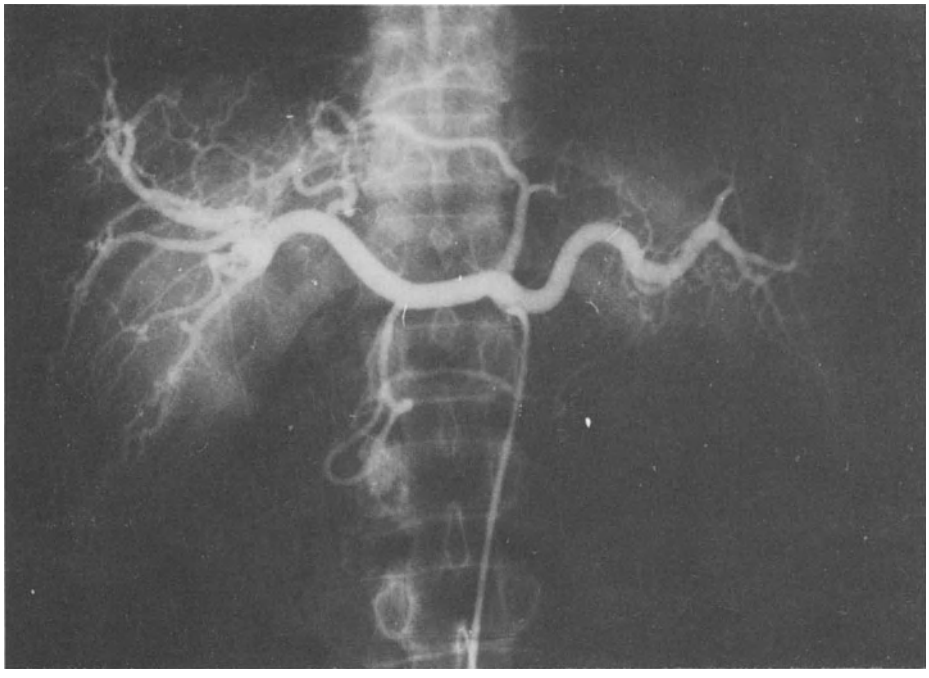
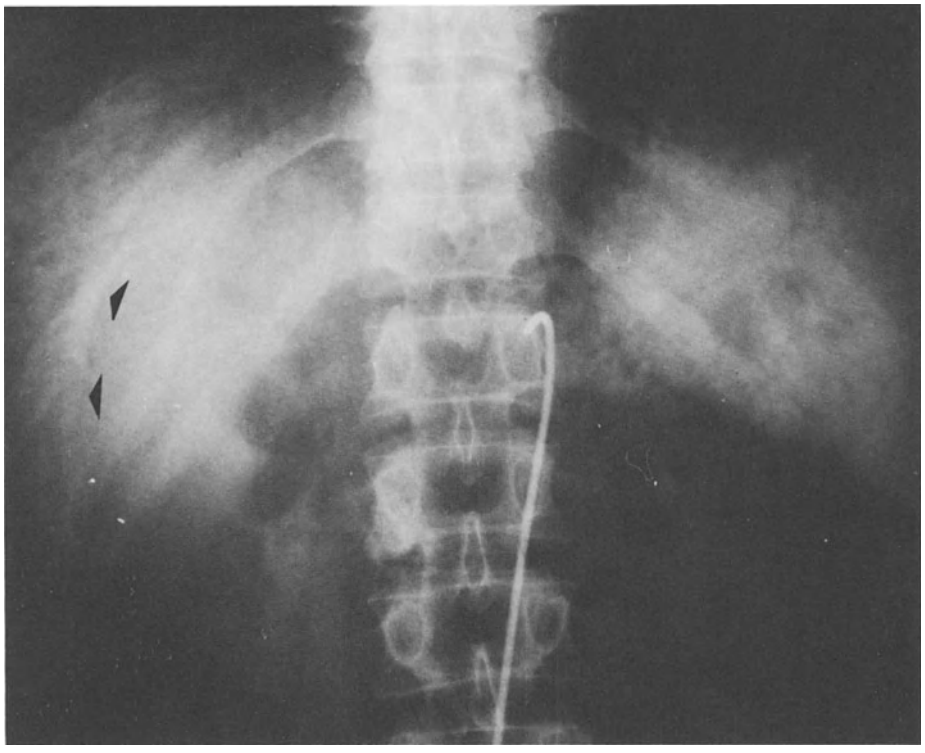


Fig. 22. Pancreatitis with portal vein thrombosis in a 45-year-old male.
Widened intrahepatic arterial branches and dense arterial hepatography resulting in good opacification of right hepatic vein 12 seconds following selective injection of contrast medium in hepatic artery.



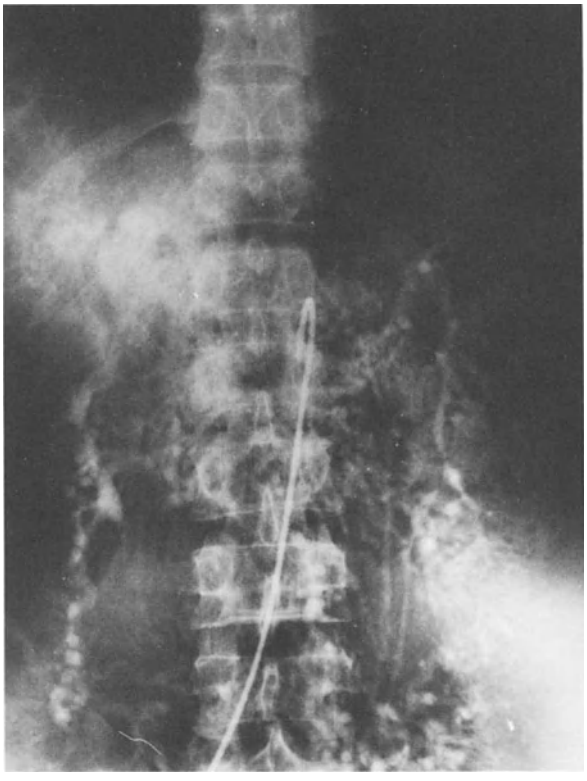
23a

Fig. 23. Chronic pancreatitis with portal vein thrombosis in a 35-year-old male (11–3–83).
23a: Normal liver with markedly dilated hepatic artery. Moderate enlargement of the spleen (15 cm) with relatively thin intrasplenic arterial branches.

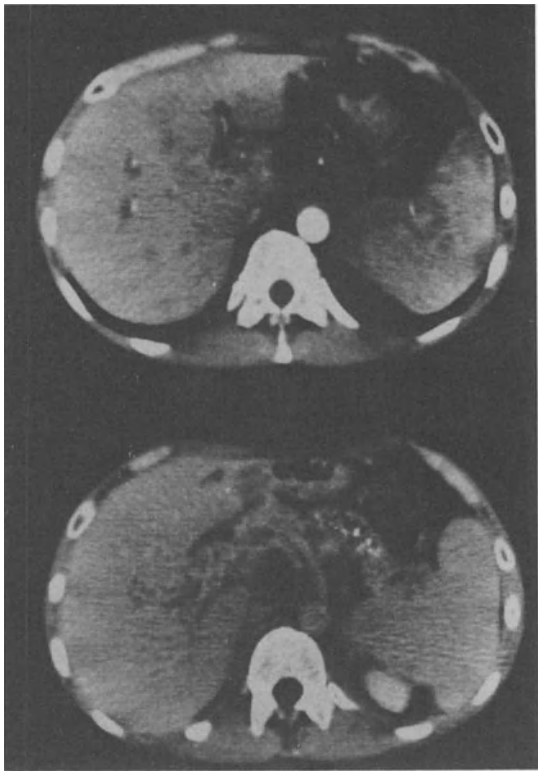


23b

23b: Splenic vein is not opacified while hepatic veins are faintly visualized.



23c



23d

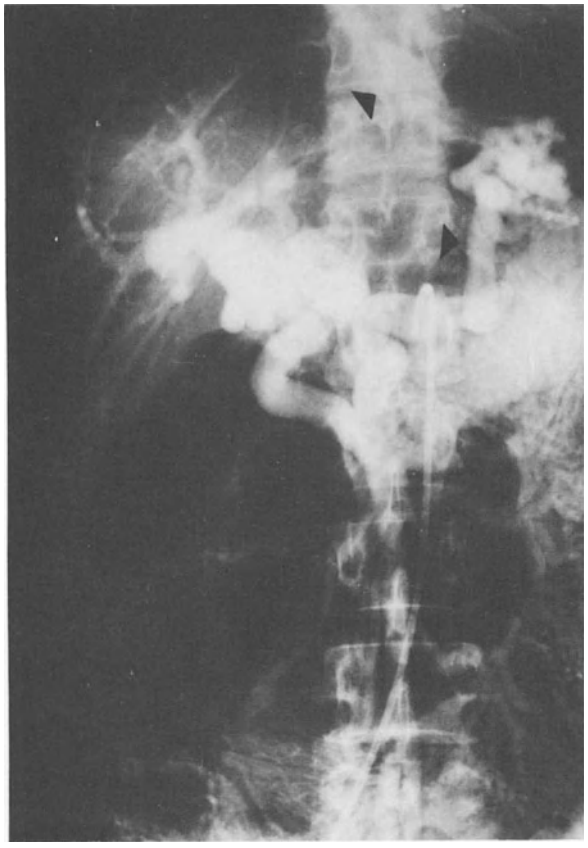


Fig. 24. Congenital obstruction of the portal vein in a 24-year-old female (9–8–77) treated by splenorenal anastomosis when she was 8 years old.
Angiographic checking for persistent esophageal varices without recurrent bleeding.
Portal vein obstruction well compensated by the development of wide hepatopetal collateral veins in the hepatic pedicule allowing for a good portal inflow.
Hepatofugal flow through left gastric vein and varices (↑). The shunt is no longer patent.

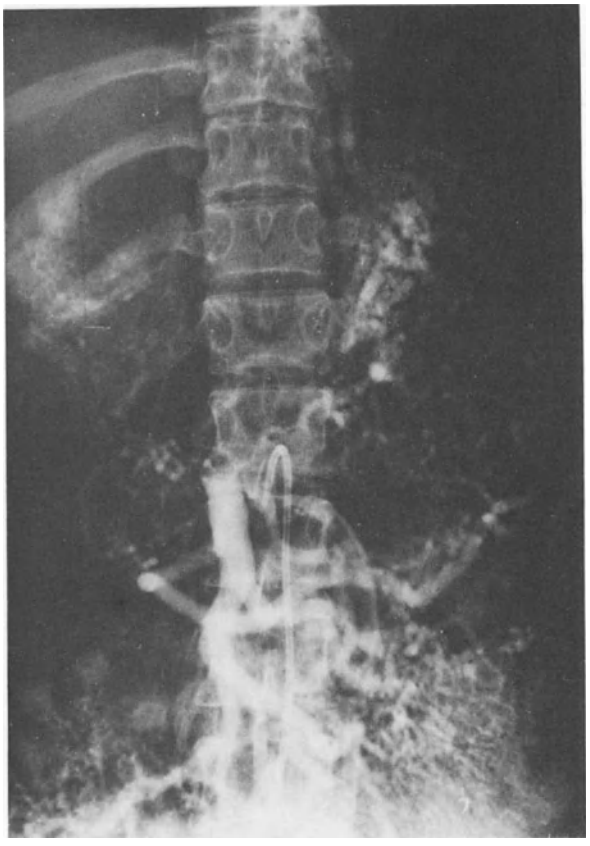
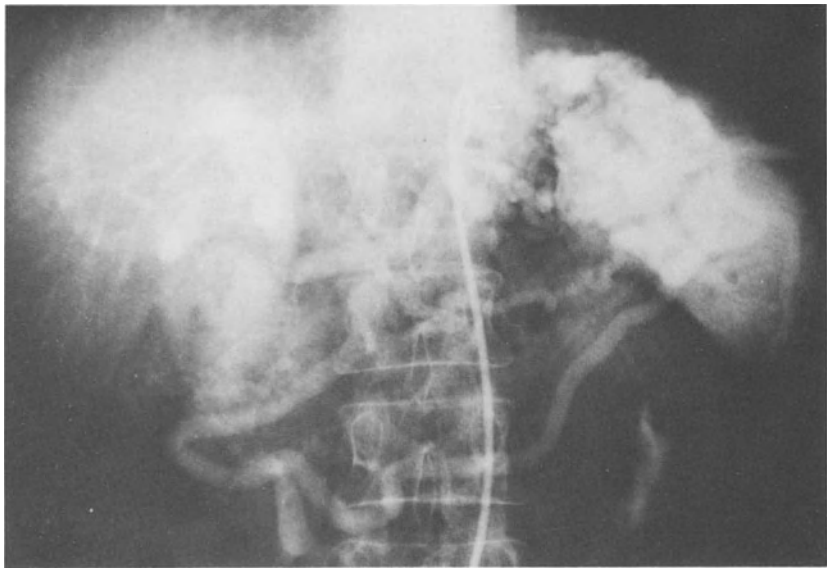


Fig. 25. Portal vein thrombosis associated with a primary Budd-Chiari syndrome in a 25-year-old female (23–8–76).
Complete thrombosis of the portal vein with only hepatofugal flow through gastro-epiploic vein and gastro-esophageal varices which signifies the post-sinusoidal obstruction.

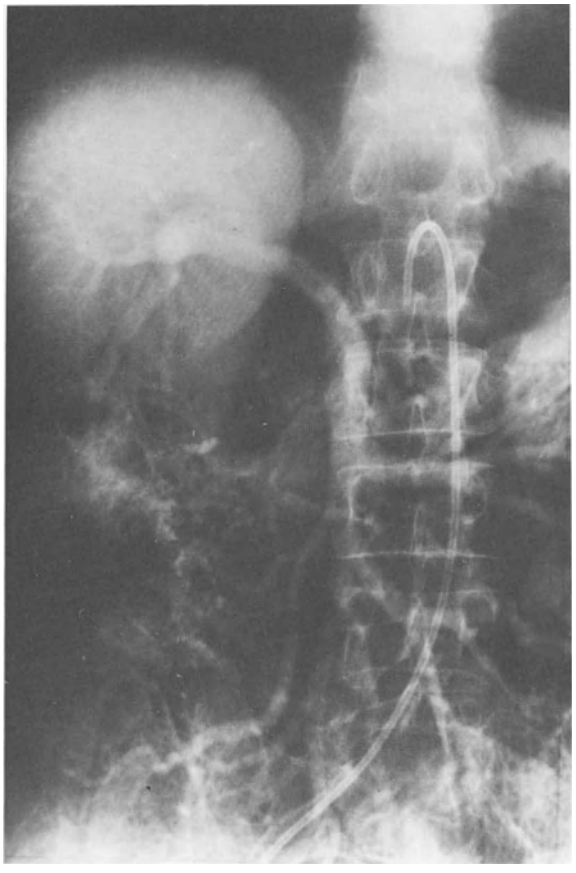
←

23c: Venous phase of S.M.A. shows complete obstruction of the mesenterico-portal axis with a poor hepatopetal portal flow.
23d: Angio C.T. (23–3–83): 15 sec after bolus injection, two dilated intrahepatic arterial branches are visualized in the upper part of the right lobe. The unopacified portal branches are seen as small lucent areas alongside the arterial branches (left gastric artery and left branch of the hepatic artery are also opacified as compared to arteriography).
40 seconds later the portal vein is not opacified and a few irregular channels within the liver hilum correspond to the hepatopetal collateral circulation.



26a

Fig. 26. Chronic pancreatitis with splenic vein obstruction in a 44-year-old male.
Patient referred for epigastric pain and upper gastrointestinal bleeding (11–3–76).
26a: Venous phase of coeliac arteriography: Splenic vein obstruction well compensated by hepatopetal collateral flow through widened gastroepiploic veins running along the greater curvature of the stomach and the left gastric and pyloric veins running along the lesser curvature as well.



26b

26b: Venous phase of S.M.A.: Normal mesenterico-portal axis and normal portal hepatography.

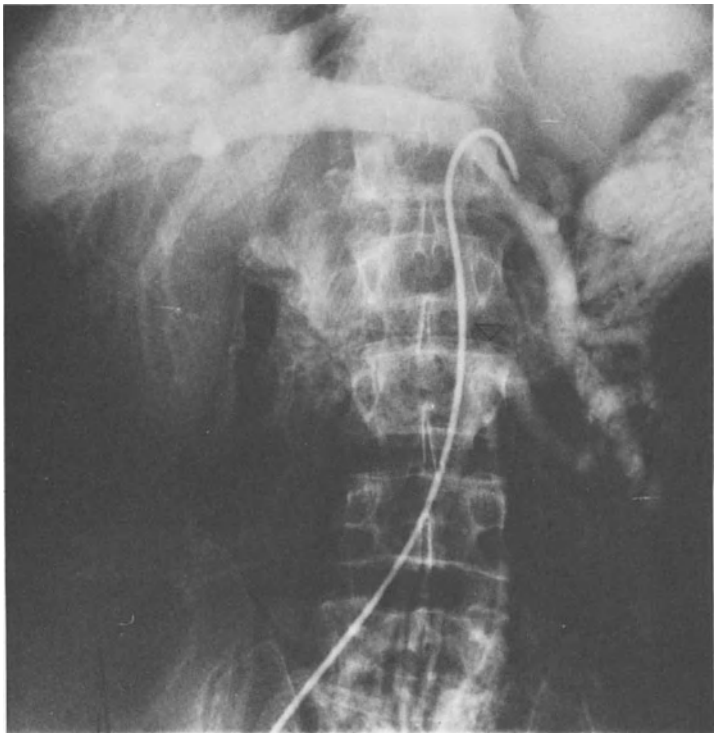
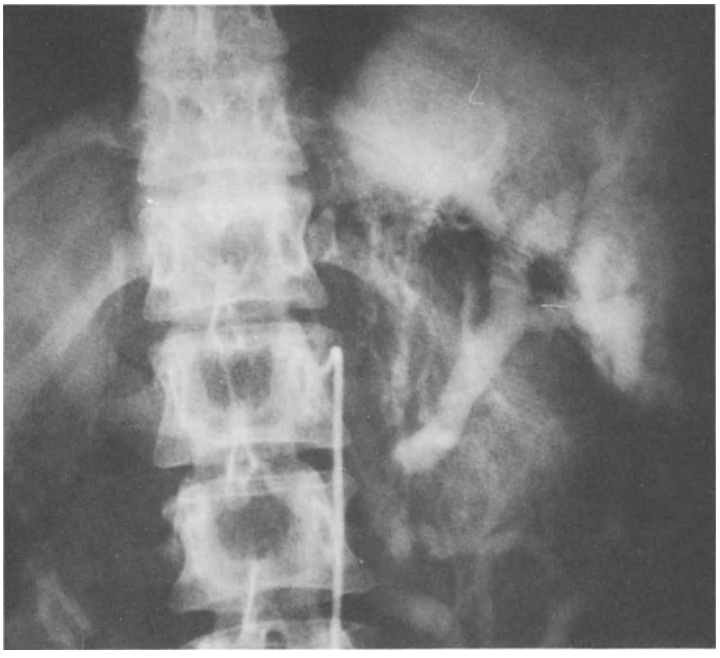
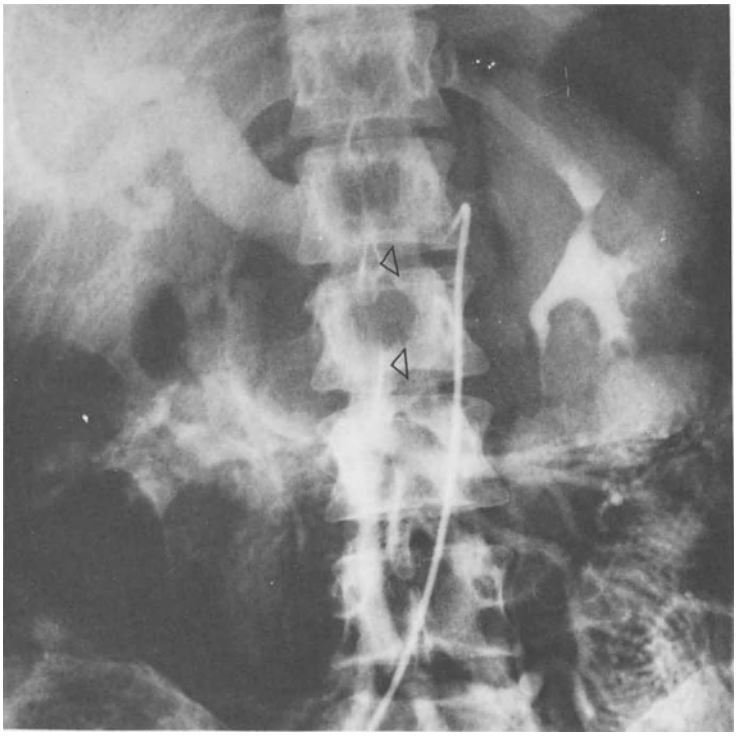


Fig. 27. Pancreatic pseudo-cyst involving the pancreatic head and the processus uncinatus as well in a 39-year-old male (3–11–76). Venous phase of S.M.A.: complete obstruction of the superior mesenteric vein (↓) with a good hepatopetal collateral flow through the markedly dilated first jejunal vein. Splenic vein was normal on the venous phase of coeliac arteriography.



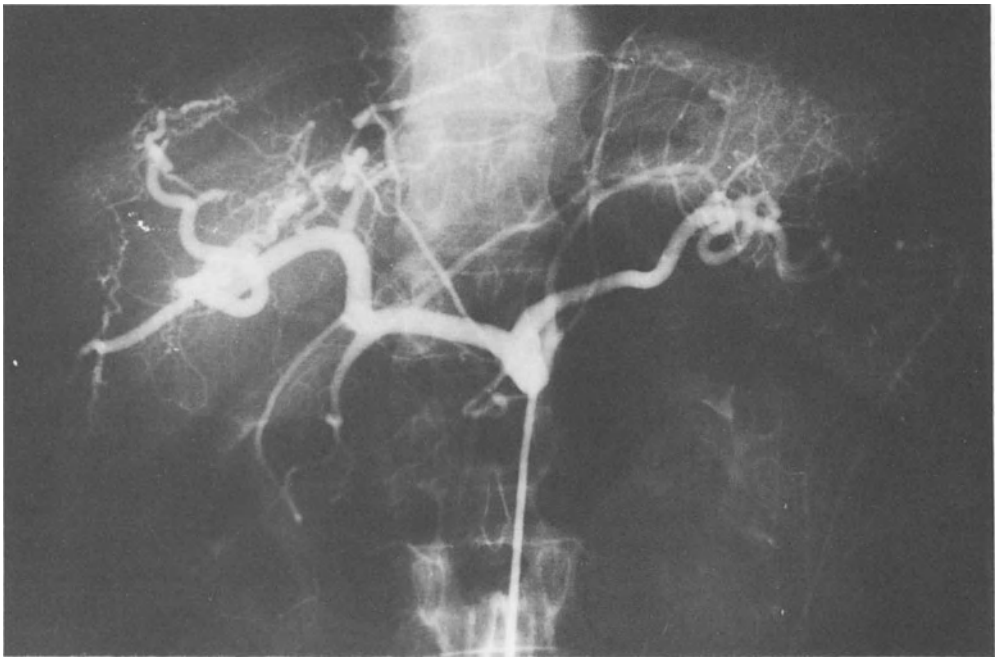
28a

Fig. 28. Obstruction of the spleno-mesenterico-portal confluence by a pancreatic pseudo-cyst involving the pancreatic isthmus in a 43-year-old female (26–4–79).
28a: Obstruction of the distal part of the splenic vein associated with a poor hepatopetal flow and a minimal splenomegaly (14.5 cm).



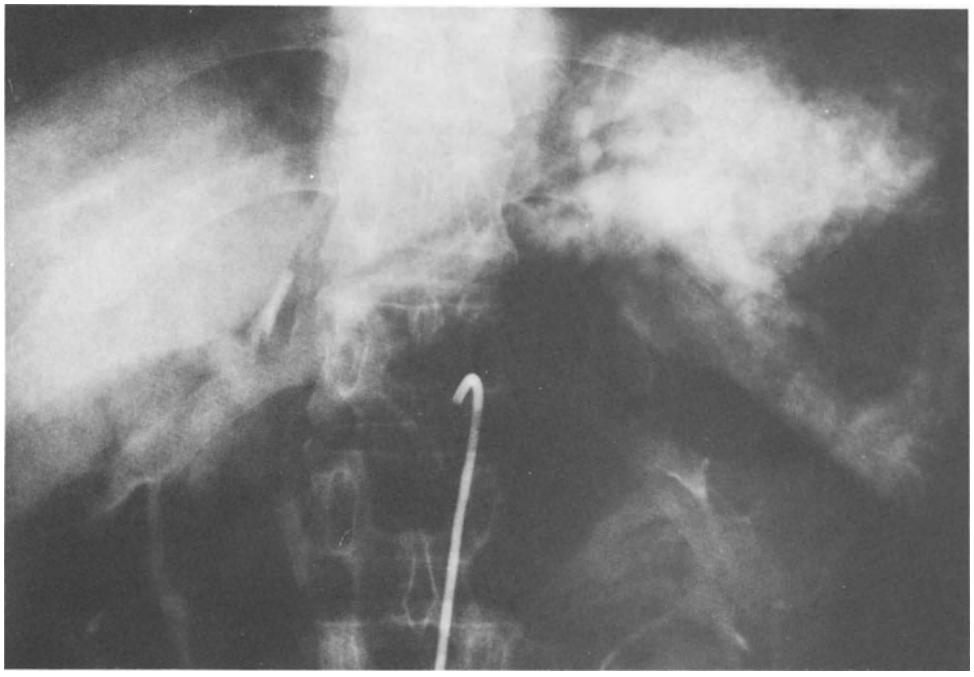
28b

28b: Narrowing of the mesenterico-portal confluence (↑) allowing, however, for a food filling of the portal vein.



29a

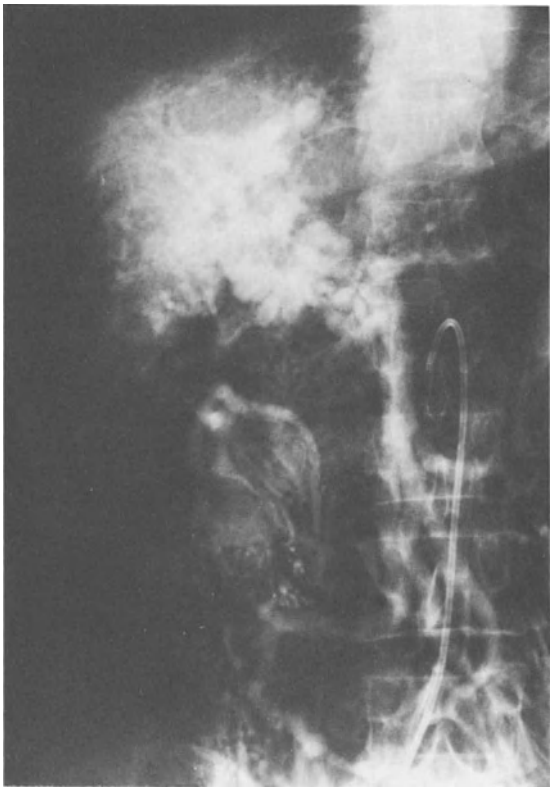
Fig. 29. Splenic and portal vein thrombosis associated with chronic pancreatitis (1–8–75).
29a: The markedly dilated hepatic artery with an otherwise normal liver suggests a severe obstruction to the portal flow of the prehepatic type. Minimal splenomegaly (14.5 cm) with very thin intrasplenic arterial branches signifies the reduced splenic blood flow.



29b

29b: Obstruction of the splenic vein with very poor venous collateral flow.

29c



29c: Venous phase of S.M.A. shows complete obstruction of the portal vein with numerous and tortuous hepatopetal collateral channels in the hepatic pedicle.

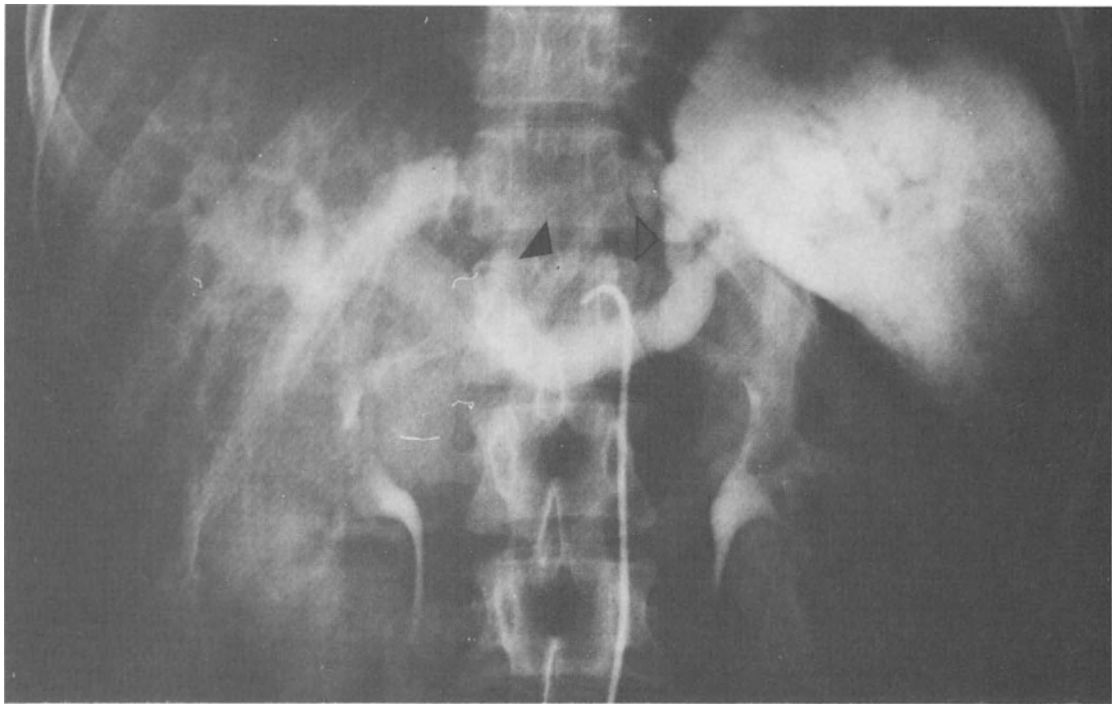


Fig. 30. Tuberculosis of the spleen with obstruction of the splenic vein due to tuberculous lymph modes in a 23-year-old female. Patient referred for bleeding from gastric varices (24–9–79).
Short segmental obstruction of the splenic vein in the splenic hilum well compensated by hepatopetal collateral flow through gastric varices (△), widened short gastric veins and left gastric vein (▲).

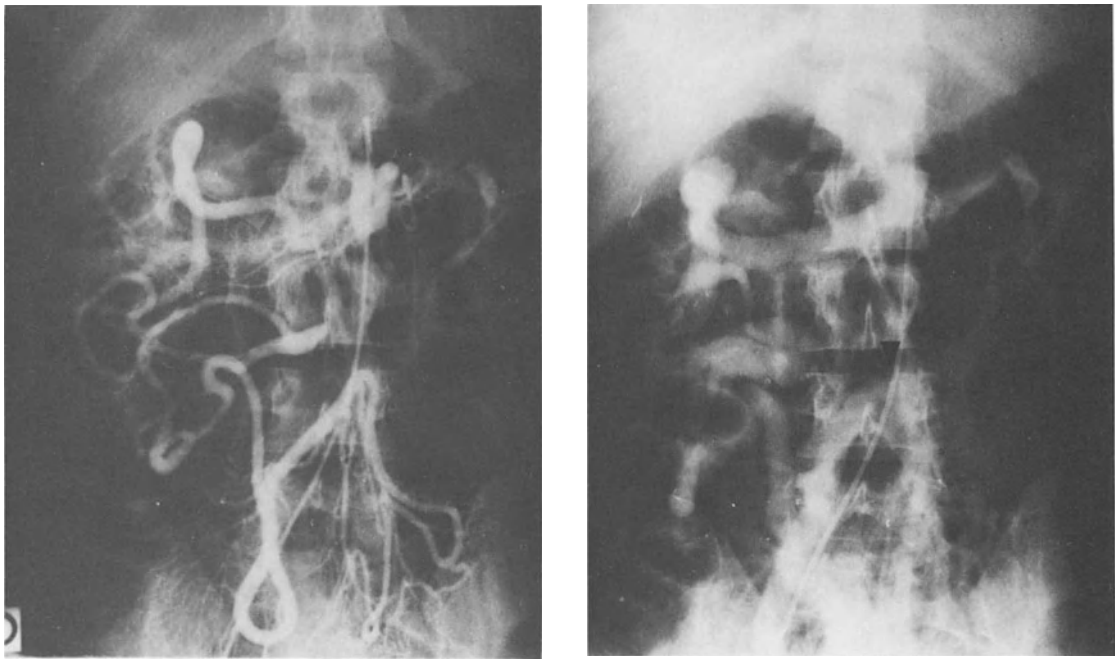
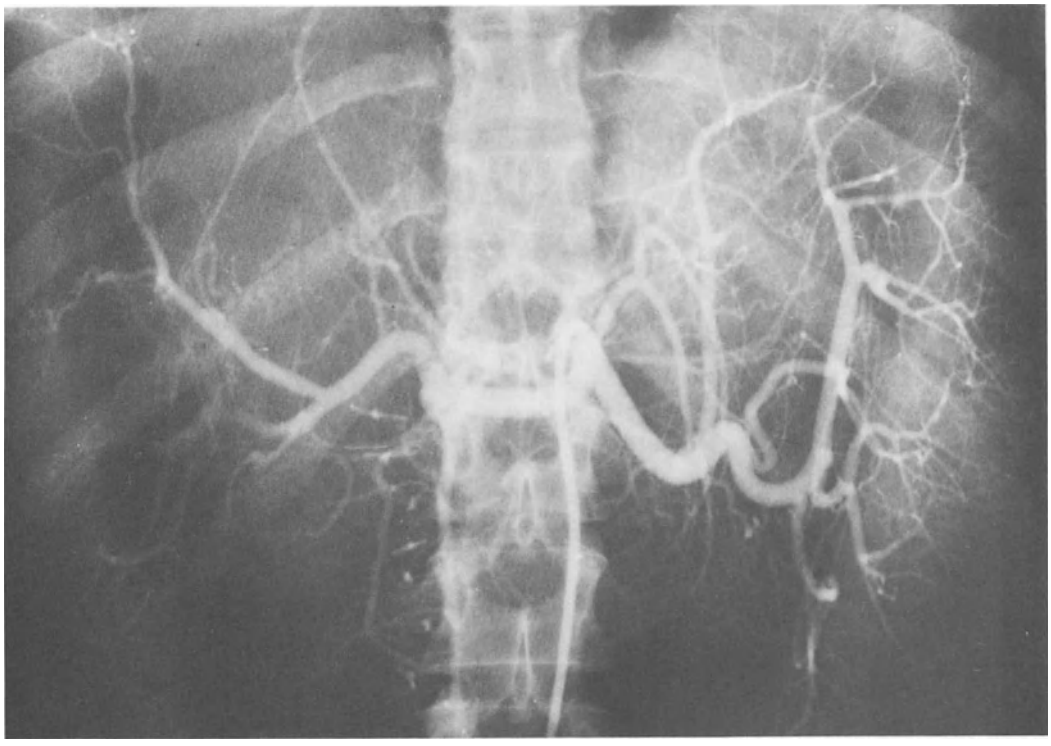


Fig. 31. Chronic mid-gut volvulus with occlusion of the superior mesenteric vein responsible for bleeding intestinal varices in a 25-year-old male (7–6–79).
Complete obstruction of the superior mesenteric artery (31a) and the superior mesenteric vein (31b) (↑) with collateral circulation through right colic arterial and venous arcades.

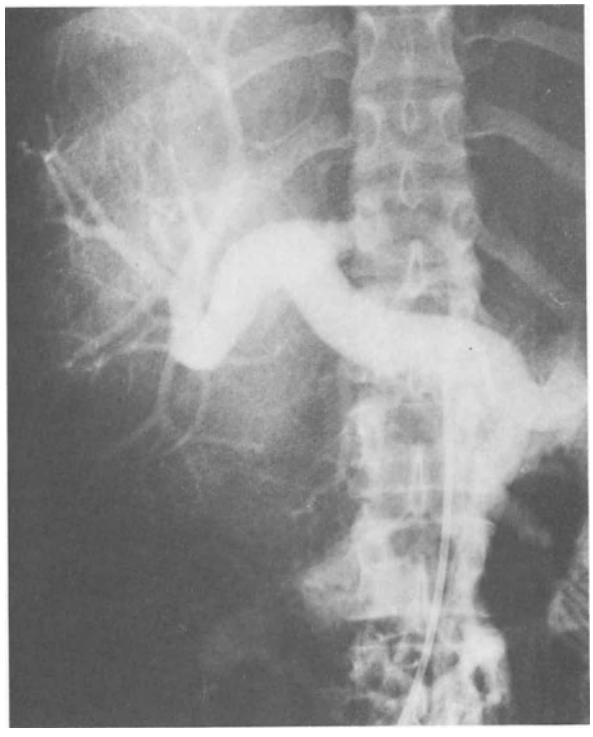


32a

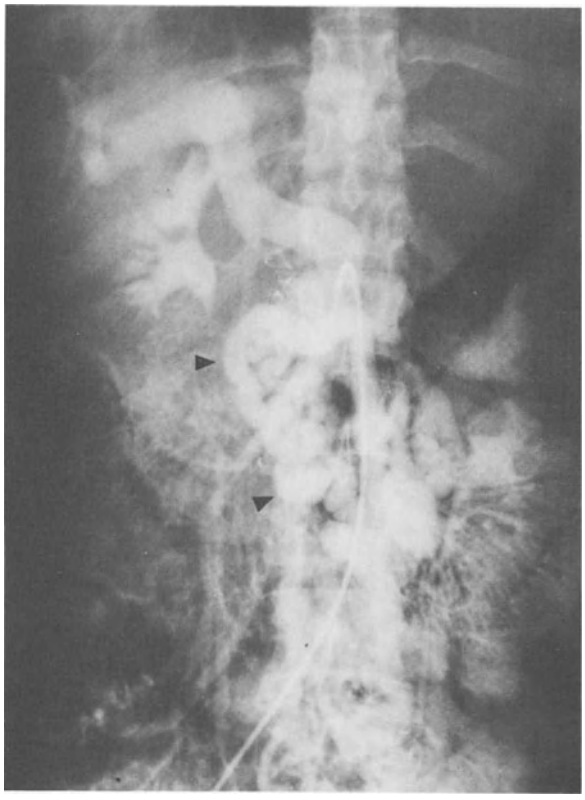
Fig. 32. Congenital fibro-angio-adenonatosis of the liver associated with a choledoceal cyst treated by biliary-digestive anastomosis in a 25-year-old female (6–5–75).

32a: Coeliac arteriography (6–5–75). Diffuse hepatomegaly and marked enlargement of the spleen (22 cm) which signifies the long-standing and slowly progressive portal hypertension.

32b

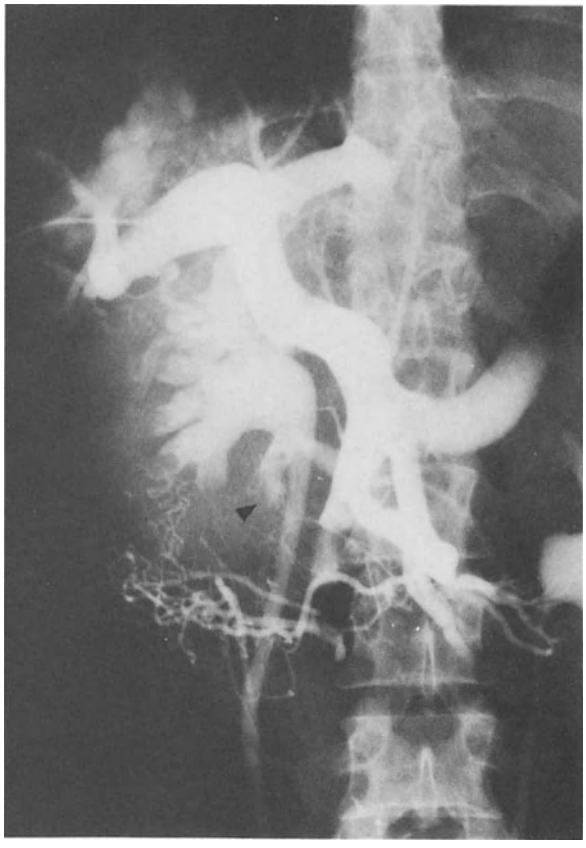


32b: Venous phase of S.M.A. (6–5–75). The portal opacification is relatively good in the right liver lobe while very poor in the left one.



32c

32c: Emergency arteriography performed 6 years later (7–1–81) for massive bleeding from duodenal origin as demonstrated by endoscopy. Venous phase of S.M.A. shows atrophy of the liver, decreased hepatopetal flow and retrograde opacification of large duodenal varices responsible for the bleeding (▲).



32d

32d: Transhepatic obliteration of the duodenal varices with Bucrylat permitted the bleeding to be immediately controlled. Obstruction of the pedicle feeding the varices is demonstrated (▲).

5. DIFFUSE POST-SINUSOIDAL OBSTRUCTIONS

I. Proper arteriographic signs

In case of intrahepatic obstruction, hepatic venography is the one method allowing the presinusoidal or post-sinusoidal site of the obstacle to be clearly recognized. In case of presinusoidal obstruction hepatic venography and wedged hepatic vein pressure are normal. On the other hand, an increased wedged hepatic vein pressure with heterogenous sinusoidal filling, narrowing and distortion of small hepatic venules on hepatic venography are typical of a post-sinusoidal obstacle.

From a hemodynamical point of view, however, two specific findings of post-sinusoidal obstructions may be documented on arterioportography.

- The essentially hepatofugal collateral venous circulation without any hepatopetal collateral flow is a common finding in every case (Fig. 25).
- The reversal of intrahepatic portal flow is an inconstant finding.

1. The porto-systemic hepatofugal circulation

The development of porto-systemic collateral anastomosis probably depends on the degree and the duration of portal hypertension as well as the anatomical possibilities in every particular case. Thus, variable angiographic patterns may be encountered.

If portal hypertension occurs in a patient where the anatomical pathways of such porto-systemic anastomosis are already present, the hepatofugal collateral flow may be visualized at an early stage of the disease even though the obstruction to the hepatopetal portal flow is minimal (Fig. 33). These anastomoses develop pregressively during the course of the disease and the more severe the obstruction to the portal inflow the higher the hepatofugal flow rate through the porto-systemic anastomosis.

On the other hand, the level of the portal pressure depends, to a certain extent, on the more or less important development of these hepatofugal collateral pathways. Thus, the higher the hepatofugal flow rate, the lower the portal hypertension, even though the degree of the obstruction is the same.

By using percutaneous transhepatic portography certain authors have noticed that the level of the portal pressure did not appear to correlate well with the size of the hepatofugal collateral pathways (42).

This is in fact all the less surprising as the portal pressure depends also on the degree of the obstruction and the subsequent decrease of the hepatopetal portal flow which might be very different from one case to another. Besides, transhepatic portography is certainly a very good method for demonstrating the various collateral pathways from a morphological point of view, depending on the positioning of the catheter tip; wheras it is certainly less reliable from a hemodynamical point of view. It is quite clear that a high flow rate spontaneous porto-caval anastomosis enables the portal hypertension to remain moderate even though a very tight obstruction results in a complete interruption of the hepatopetal portal flow. Owing to this uncommon hemodynamic condition, some patients will never have any digestive bleeding whereas the liver disease may remain clinically unapparent until the late stage of atrophic cirrhosis associated with a severe hepatic failure and encephalopathy is reached. Furthermore, if the liver disease responsible for the obstruction to the portal inflow is not associated with hepatic failure, the portal hypertension may remain totally unknown in spite of a totally hepatofugal portal flow (Fig. 38).

On the contrary, when only a few collateral pathways allow a poor hepatofugal flow a higher portal hypertension will result either in more severe gastro-intestinal hemorrhage or ascitis as it is usual in the Budd-Chiari syndrome. Another example of this hemodynamical condition is the pronounced

and the recurrent ascitis occurring in cirrhosis after transhepatic embolization of large and high flow rate esophageal varices and resulting from the substantial increase of the portal pressure. In fact, the hepatofugal collateral flow is a common phenomenon in every case of post-sinusoidal obstruction whatever its various anatomical pathways.

Therefore, angiography must be able to demonstrate whether such a hepatofugal collateral flow exists or not. But, from a diagnostic point of view, it is not necessary to document as completely as possible the morphological pattern of these anatomical pathways, whereas it is more useful to evaluate its hemodynamical significance, that is to say the level of the hepatofugal flow rate.

Opacification of the porto-caval anastomosis depends on the concentration of the contrast medium in the portal vein and the hepatofugal flow rate through these collateral pathways. The higher the shunt flow rate, the larger the amount of contrast medium flowing through it. Conversely, the lower the flow rate, the smaller the amount of contrast medium and the more weakened will be the opacification due to dilution by the non-opacified blood. This is why some low flow rate porto-systemic anastomoses cannot be opacified even though the contrast medium is injected directly into the portal vein as when using transhepatic portography. It is a common situation when portography is performed for recurrent bleeding after transhepatic embolization of esophageal varices.

Because the recurrent varices are usually small and poorly supplied with blood from short gastric veins, their opacification may require a selective catheterisation of the feeding vein whereas they are not visualized on the portography. Nevertheless, the angiographic methods, using a direct opacification of the portal system (splenoportography, ombilico-portography, transhepatic portography) are obviously the most efficient to document the porto-systemic collateral circulation whatever its hepatofugal flow rate. Thus, they are very useful from a morphological point of view. However, they do not provide as reliable information about the hemodynamics of the portal flow, as they are not physiological methods.

As a matter of fact, the contrast medium injection rate into the portal vein may probably give rise to transient hemodynamical changes. In addition, the portal flow rate and direction may not be the same in different areas of the portal system, so that the angiographic pattern is likely to depend on the precise site where the contrast medium is injected.

On other hand, arterioportography is a more physiological method, allowing for a good visualization of the whole mesenterico portal axis which is the main afferent way to the liver from a functional point of view. The contrast medium is introduced into the superior mesenteric artery with a low injection rate (6 ml/sec), which is probably nearly identical to the physiological blood flow rate. Thus, it is likely that the injection of contrast medium without any drug-induced vasodilatation does not result in significant changes of the normal hemodynamics.

Once the dye has flowed through the capillary bed to fill in the superior mesenteric vein, the subsequent opacification of the portal venous system is likely to be a reliable result of the hemodynamics of the portal flow.

Therefore, the venous phase of the superior mesenteric arteriography may be considered to be the best angiographic method to document the portal blood flow abnormalities, allowing for the portal inflow to be evaluated as compared to the hepatofugal portal flow.

It may be though that the higher the flow rate through a porto-systemic anastomosis is the more dilated and the better visualized on a greater length the collateral venous pathway is, since the contrast medium is all the less weakened by the non-opacified blood. For this reason, some spontaneous high flow rate porto-systemic anastomosis results in a dense opacification of the inferior vena cava itself (Figs. 35, 36, 37, 38).

Actually, the low flow rate hepatofugal collateral veins are only faintly opacified and may be not visualized. The arteriportography may be less efficient than other angiographic methods from a morphological point of view, while more reliable from a physiological one (Fig. 34).

The hepatofugal collateral circulation may flow through many different anatomical pathways allowing for a more or less important flow rate. In practice, however, they can be divided into two different groups according to their frequency and their blood flow rate possibilities. The first group corresponds to the common anastomosis whose

flow rate is usually low or moderate. The second group is made up of less common and possibly high flow rate anastomoses.

a) The common porto-systemic anastomoses with a low or moderate hepatofugal flow rate

The gastro-esophageal anastomosis. The gastro-esophageal anastomosis through which the blood drains from the left gastric vein and the short gastric veins toward the azygos vein and the superior vena cava have utmost clinical importance as they are responsible for bleeding from esophageal varices.

However, these porto-systemic anastomoses have little hemodynamical significance as the hepatofugal flow rate through the varices is usually relatively low. This may be demonstrated by performing transhepatic embolization of esophageal varices as, in most cases, the portal pressure increase resulting from varices obliteration is only moderate: 3 to 5 mm Hg.

Due to this low hepatofugal flow rate arteriography allows only for a faint or incomplete visualization of these gastro-esophageal collateral pathways in many cases. Therefore, the development of esophageal varices as evidenced by endoscopy or baryum swallowing, which results from the blood stasis within the submucosal venous plexus, does not correlate with their arteriographic opacification which depends mainly on the hepatofugal flow rate through these varices (Fig. 34).

Hemorrhoidal anastomosis. The development of communications between inferior mesenteric vein and inferior vena cava through the rectal veins is a common finding in portal hypertension but these anastomoses have no hemodynamical significance. However, retrograde opacification of the inferior mesenteric vein is one of the commonest angiographic feature to account for the hepatofugal flow in portal hypertension.

Retroperitoneal anastomoses. Porto-caval anastomoses may occur wherever the gastro-intestinal tract lies in close contact with veins that are tributaries to the inferior vena cava, i.e. in the retroperitoneal space.

In normal conditions, such anastomoses occur mainly in the anterior para-renal space where the intestinal veins draining the blood from the right and left colon may drain towards the inferior vena cava either through the lumbar veins (Retzius anastomosis) or the perirenal and gonadal veins (Fig. 33). Similarly the splenic and gastric venous blood may drain through diaphragmatic and retroperitoneal veins towards the inferior vena cava, through the gastro-phrenic and spleno-renal ligaments.

Unusual porto-systemic communications may incidentally occur through abnormal adhesions resulting from previous abdominal surgery. Although the hepatofugal flow rate through these anastomoses is usually low, it may induce the development of varices in unexpected sites. An example of this phenomenon is demonstrated in patients treated for biliary obstruction by a surgical anastomosis between the common bile duct and a jejunal loop. On account of this portal hypertension may result in either duodenal or jejunal varices leading to lower gastro-intestinal bleeding (Fig. 32) (43).

Although an exceptional finding, hematuria from bladder varices may probably result from post-surgical adhesions between the ileal loops and the urinary bladder. Finally gastro-esophageal, hemorrhoidal or intestinal anastomoses usually have little hemodynamical value but great clinical importance due to their potential for gastro-intestinal hemorrhage.

b) Spontaneous and potentially significant porto-systemic shunts

Some portocaval anastomoses are less common as they probably result from pre-existing anatomical channels such as embryonic ones which widen and elongate on account of portal hypertension, thus allowing for a relatively high hepatofugal flow rate. They are mainly the umbilical or paraumbilical vein, the spleno-renal collateral and the meso-gonadal anastomoses. As a matter of fact, these porto-systemic anastomoses are not so rare when associated with a low or moderate hepatofugal flow rate. They are then identical to the common porto-systemic anastomoses from a hemodynamical point of view and do not deserve special mention.

In a few cases, however, the hepatofugal blood

flow rate through these markedly widened collateral veins is so high that the portal pressure is either only slightly increased or even normal or near normal, in spite of a severe obstruction to the portal inflow which is markedly reduced. As a result, spontaneous porto-systemic anastomosis may hinder the forming of esophageal varices and prevent bleeding as well as ascitis, so that the disease may be clinically inconspicuous, mainly in non-cirrhotic patients, (Fig. 38).

Some cases of cirrhosis may remain unknown for many years until the liver is atrophic enough to give rise to severe hepatic failure at a late stage of the disease. However, these high flow rate porto-caval anastomoses often induce some peculiar clinical or biological disorders such as diabetes, hemochromatosis, peptic disease, and encephalopathy.

Umbilical and paraumbilical anastomosis is the most common. It arises from the left portal vein main branch, then runs to the umbilical area and joins the epigastric veins to communicate with the inferior or occasionally superior vena cava. The higher the flow rate through this anastomosis, the more widened the left portal vein branch and the umbilical vein and the denser their opacification. As a result, the inferior vena cava itself may be clearly visualized a few seconds later (Fig. 35).

In some cases the splenic vein is also seen draining through this umbilical anatomosis so that the inferior vena cava may be visualized on the venous phase of splenic arteriography as well. However, the opacification of the inferior vena cava is always weakened by the non opacified blood flow from the superior mesenteric vein.

A relative value of the increased hepatofugal flow rate can be performed in relation with the decreased hepatopetal one. The relative value of the hepatofugal and hepatopetal flow rate may be evaluated after the widening in diameter of the left portal vein branch contrasting with the narrowing of the right one.

Spleno-renal and gastro-renal anastomoses usually link the splenic vein with the left renal vein via the left adrenal vein through the short gastric veins and the left inferior phrenic vein. The communicating channels may be relatively short in a few rare cases but they usually run through a long and tortuous network of dilated gastric veins, often responsible for gastric varices, whereas there are no or only few esophageal varices.

In spite of the risk of severe bleeding associated with gastric varices, this spontaneous spleno-renal anastomosis sometimes permits the level of the portal pressure to be lowered and the complications of portal hypertension to be prevented for a time. In the same way a dense and early filling with contrast medium of the left renal vein and inferior vena cava is well documented on the venous phase of the superior mesenteric arteriography and the coeliac arteriography as well.

Unlike this collateral venous pathway the portal vein and its intrahepatic branches are narrowed and faintly opacified which accounts for a markedly decreased hepatopetal portal flow as compared to the high hepatofugal flow rate. Coeliac arteriography clearly demonstrates this very peculiar hemodynamical status by documenting a markedly widened hepatic artery, which means that hepatopetal portal flow is very poor. It also elicits a normal or near normal splenic pattern accounting for a normal or near normal portal pressure.

Thus the utterly different significance of these two angiographic signs, i.e. the hepatic arterial and splenic changes, is clearly pointed out, together with the possibility for a severe intrahepatic obstruction to be associated with slight portal hypertension. In this circumstance, the unusual discrepancy between both signs enables one to anticipate the existence of a high flow rate porto-systemic anastomosis on the basis of the arterial phase of the coeliac arteriography only (Fig. 36).

Meso-gonadal anastomoses are less frequent but may sometimes carry a high hepatofugal blood flow rate (43). They usually link the superior mesenteric vein with the right gonadal vein through the dilated and elongated ileo-colic veins forming a tortuous venous network in projection of the right iliac crest (Fig. 33). Less commonly this anastomosis links the inferior mesenteric vein to the left gonadal vein.

The gonadal vein may be dilated enough to allow for a high hepatofugal flow rate, in such a way that the inferior vena cava itself is well documented on the late venous phase of superior mesenteric arteriography (Figs. 37, 43). In addition, the ileo-colic veins may also drain towards the perirenal

veins which are always connected with the gonadal veins (Lejars's anastomosis) (Fig. 37). Other anatomical pathways than gonadal and perirenal veins may allow the mesenteric blood to drain toward inferior vena cava, although the former are by far the most commonly encountered.

In a probably very unusual case, a wide and relatively direct retroperitoneal anastomosis was seen to unite the superior mesenteric vein to the left renal vein, probably through the middle colic vein. In this non-cirrhotic patient, portal hypertension was inconspicuous in spite of a completely interrupted hepatopetal portal flow (Fig. 38). Although relatively rarely encountered, this spontaneous porto-caval anastomosis may be clinically interesting as it is usually not associated with the formation of intestinal varices and consequently with potential risk of bleeding.

Finally, the clinical course of the disease in most patients with a high flow rate porto-caval anastomosis proves the efficiency of such spontaneous shunts to prevent or delay the complications associated with portal hypertension. This is why the disease may long remain clinically unrecognized.

However, such porto-caval anastomoses are relatively rare and although they allow the level of the portal pressure to be reduced, they are unable to lower it to normal level in any case. As a matter of fact, the hepatofugal collateral flow should be considered as resulting mainly from the decreased hepatopetal portal flow which is closely correlated to the increased intra-hepatic resistances. It is commonly found that a high hepatofugal flow rate is always associated with a markedly decreased or even completely interrupted hepatopetal portal flow.

Whatever the degree of portal pressure lowering they are responsible for, these overdeveloped porto-systemic anastomoses seem to be the result of a severe and probably long-standing obstacle to the hepatopetal portal flow, which is the primary causative factor from a physiopathological point of view (42).

2. Reversal of intrahepatic portal flow

Intrahepatic portal flow reversal is a relatively uncommon hemodynamic abnormality resulting from a post-sinusoidal obstruction. It was initially documented by hepatic venography in cirrhotic patients who had undergone surgical porto-caval anastomoses (44). However, reversal of the intrahepatic portal flow may occur spontaneously in any case of post-sinusoidal obstruction, whatever its cause and its site.

It is now well known that this phenomenon may be evidenced by arterioportography in some cases of cirrhosis (15, 18, 45, 46) and Budd-Chiari syndrome as well (47, 48). In addition it is also the main sign allowing for a segmental intrahepatic obstruction without portal hypertension to be recognized by arterioportography (49, 50) as it will be discussed later (Chapter VIII).

The occurrence of reversed intrahepatic portal flow in the right lobe of the liver after surgical ligation of a ruptured right hepatic vein in a case of abdominal contusion serves to demonstrate that hepatic vein occlusion results in reversal of portal flow in the corresponding area of the liver (51). This reversed portal flow is due to an increased arterial flow resulting from the interruption of the hepatopetal portal flow and probably the development of intrahepatic arterio-portal shunting as well.

As a matter of fact, the tighter the obstruction, the more slowed the portal inflow and the higher the arterial flow rate. Owing to the progressive increase of intrahepatic resistances, and the development of hepatofugal porto-systemic anastomoses, a more important part of the arterial blood feeding the liver will flow backward through the intrahepatic portal branches to drain towards the porto-caval anastomoses.

Thus the cirrhotic liver which was a four-way circulatory system (two afferent ways the portal vein and the hepatic artery and two efferent ways: the hepatic veins and the porto-systemic anastomosis) (Fig. 2) becomes a two-way system (one afferent way: the hepatic artery and one efferent way the portal vein) when the hepatic venous bed obstruction is severe enough (Fig. 39).

The angiographic demonstration of this reversed intrahepatic portal flow relies on the visualization of a retrograde opacification of the intrahepatic portal branches through the injection of contrast medium into the hepatic artery, i.e. an arterioportal reflux. It will be all the more easily evidenced as the arterial flow rate is higher, the intra-

hepatic arterio-portal shunting more developed and the hepatofugal portal flow rate more increased.

In fact, even though arterio-portal communications do exist in normal liver as suggested by some experimental works (Chapter 1), they could not be demonstrated by arteriography since the potential opacification of small portal branches through these shunts is immediately weakened by the thrice higher non-opacified portal blood flow. Conversely the more slowed the portal inflow, the higher the arterial flow and the more concentrated is the contrast medium injected into the hepatic artery flowing back through the intrahepatic portal veins, so that they can be visualized.

The intrahepatic arterio-portal communications which are well known to develop in cirrhosis, as demonstrated first by the experimental works of Herrick in 1907 (52), constitute a favorable condition for the reversal of intrahepatic portal flow to occur.

Many authors have confirmed such arterio-portal shunting in the cirrhotic liver on the basis of experimental (4, 5) anatomical (6, 7) and physiological works using isotopic scanning (53) or oxygen concentration measurement in the portal vein blood (54).

By using arterioportography, the demonstration of this hemodynamic abnormality is supported by three angiographic signs: one direct sign and two indirect signs:

– *The direct sign* consists of the visualization of a retrograde opacification of the intrahepatic portal branches following injection of contrast medium into the hepatic artery.

It is then better demonstrated by selective hepatic arteriography since there is no risk for this retrograde opacification of the portal branches to be confused with the antegrade one occurring normally on the venous phase of the coeliac or superior mesenteric arteriography. However, the coeliac arteriography is often reliable enough to document this angiographic anomaly as the retrograde opacification of the intrahepatic portal branches occurs at a relatively early phase (5 to 6 seconds after the injection) whereas the venous phase of coeliac arteriography is always very faint and delayed in the cases of severe post-sinusoidal obstruction. In addition, even though the spleno-portal axis is visualized on the late films of the venous phase of coeliac arteriography, it may be noticed that opacification of some intrahepatic portal branches is not only earlier but denser than the opacification of the portal stem itself, which may only result from retrograde filling through the arterial route.

Nevertheless this angiographic pattern is probably difficult to recognize and it is likely for the reversal of intrahepatic portal flow to be missed if a selective hepatic arteriography is not performed at least in cases of low intrahepatic hepatofugal flow rate, which is probably less uncommon than usually thought.

– *An increased arterial flow* in the liver area where the portal flow is seen to be reversed constitutes a common indirect sign. In cirrhosis arterial flow is usually diffusely increased, giving rise to a dense arterial hepatography often hindering the retrograde filling of small intrahepatic portal branches to be clearly visualized. As a result of this retrograde opacification of the portal stem is only visualized on the late films of selective hepatic arteriography in many cases.

– The main indirect sign is documented by the venous phase of superior mesenteric arteriography, *that is, the lack of opacification of those intrahepatic portal branches in which the blood flow is reversed.*

The contrast medium stops at the point where both opposite blood flows meet, giving rise to a functional obstruction angiographic pattern. Unlike the true anatomical obstructions, the interrupted portal veins are seen to end unsharply as the contrast medium is diluted by the non-opacified retrograde blood flow.

At any rate the retrograde opacification of those interrupted portal veins through a selective injection of contrast medium into the hepatic artery will account for the functional origin of this obstructive pattern.

It appears that in cirrhosis, the more developed the porto-systemic anastomosis, the higher is the reversed intrahepatic portal flow rate. Consequently the reversed portal flow rate remains relatively low as far as the hepatofugal flow rate through the porto-systemic collateral pathways is only moderate. Thus some intrahepatic portal branches are only opacified by selective hepatic arteriography whereas no retrograde filling of the portal stem itself is seen.

On the contrary, the portal vein is opacified in a normal way on the venous phase of superior mesenteric arteriography while the intrahepatic portal branches are only faintly or partly visualized (Fig. 40).

This angiographic pattern means that the reversal of intrahepatic portal flow involves either certain areas of the liver only or the whole liver parenchyma but with too low a hepatofugal flow rate for the hepatopetal flow to be completely interrupted in the portal vein itself. Under these circumstances a bidirectional flow could be demonstrated in the portal vein in some cases (Fig. 41). This corresponds to a partial reversal of the portal blood flow (Fig. 39a.)

Owing to the progression of the intrahepatic obstruction and the development of the porto-systemic collateral pathways, a completely hepatofugal blood flow will occur through the portal vein. Thus a retrograde filling of the portal vein itself is documented on selective hepatic arteriography whereas the venous phase of superior mesenteric arteriography demonstrates a functional obstructive pattern of this vein (Figs. 39b, 42, 43, 44).

It is interesting to note that C.T. scan allows for the reversed intrahepatic portal flow to be clearly demonstrated, owing to rapid intravenous injection of contrast medium. On the first film corresponding to the arterial phase, early opacification of the intrahepatic portal branches in which the flow is reversed is visualized and the angio C.T. pattern compares similarly to the arteriographic pattern (Fig. 45). Indeed, partial and complete reversal of the intrahepatic portal flow are only two different stages of the same hemodynamic phenomenon, which probably depends mainly on the hepatofugal flow rate through the portocaval anastomosis.

However, the reversed intrahepatic portal flow does not result from the porto-systemic anastomosis even though the hepatofugal flow rate is all the more increased as the porto-caval collateral pathways are more developed. As a matter of fact, reversal of intrahepatic portal flow may occur after a surgical end-to-side porto-caval anastomosis in which the hepatic end of the portal vein has been ligated as demonstrated by hepatic venography (55, 56).

Because the portal vein is no more patent in these cases the reversed intrahepatic portal flow drains through small porto-mesenteric hepatofugal collateral veins which develop within the hepatic pedicle. In addition, the intrahepatic portal flow may be reversed even though the porto-systemic anastomoses are poorly developed (Fig. 40). Indeed this hemodynamic abnormality results essentially from the obstruction to the portal inflow and the development of intrahepatic arterioportal shunting. In cirrhosis it seems to be the result of a severe or quickly progressive alteration of the liver parenchyma.

In our own experience, arterioportography demonstrated the intrahepatic portal flow to be reversed in 45 cirrhotic patients, i.e. in about 5% of the cases of cirrhosis studied by arteriography. One third of these patients had a completely hepatofugal portal flow (Figs. 42, 43, 44) whereas in two-thirds of the cases a partial and perhaps intermittent (57) reversal of the intrahepatic portal flow was evidenced (Fig. 40, 41, 45). Therefore this hemodynamic anomaly is a rare but not uncommon arteriographic finding in cirrhosis.

It may be a useful sign not only for the diagnosis but mainly because it accounts for the existence of severe hepatic lesions as it could be demonstrated in some cases by liver biopsy (Fig. 40). However, the reversed intrahepatic portal flow per se has not always a poor prognostic significance.

Although it is the result of a severe obstruction of the intrahepatic venous bed, it probably allows for a relatively good blood supply of the liver parenchyma to the maintained. As a matter of fact, in some cases of non-alcoholic cirrhosis and idiopathic portal fibrosis as well a complete reversal of the intrahepatic portal flow was seen to be associated with a long survival provided the liver cell function was preserved (Figs. 38, 44).

By using hepatic venography, diffuse and severe obstruction of the hepatic venous bed within the liver may be evidenced whereas the reversed intrahepatic portal flow may be documented through wedged hepatic venography. However, this method seems to be less reliable than arterioportography since it is not as physiological a method and furthermore it permits to explore only a small area of the liver parenchyma. It is probably the reason why we observed a few discrepancies between the results of both methods: hepatic ven-

ography being sometimes unable to demonstrate the reversed intrahepatic portal flow, which was, however, well documented a few days earlier by arterio-portography.

The intrahepatic arterio-portal shunting which gives rise to portal flow reversal mainly in cirrhosis must not be misinterpreted as an intrahepatic arterioportal fistula, which is a more rare vascular lesion resulting in most cases from liver trauma. In case of fistula, there is usually only one arterioportal communication whose opacification occurs very early, between one to two seconds after contrast medium injection. The communication itself is visualized as well as the draining vein running alongside the corresponding arterial branch.

Unlike the fistula, the arterio-portal communications associated with the reversal of the intrahepatic portal flow are multiple and more or less diffusely located within the liver. Their opacification is relatively delayed, occurring between 4 to 7 seconds after contrast medium injection, following a dense and more or less heterogeneous arterial hepatography. For this reason neither the arterioportal communication itself nor the small draining vein are visualized in most cases whereas only the retrograde filling of either some segmental intrahepatic portal branches or the portal vein itself is demonstrated.

Therefore, the angiographic patterns of the intrahepatic arterioportal fistulas and the reversal of the portal flow are quite different and should not be confused. Indeed, in the case of portal flow reversal the communications between the hepatic arteries and the portal veins are probably much thinner and more distally located than in case of fistula. However, in some cases of cirrhosis or idiopathic portal fibrosis arterioportal communications associated with portal flow reversal may liken arterioportal fistula since they are probably more proximally located. Although rarely encountered, these cases raise the question as to whether an intrahepatic arterio-portal fistula may result from the cirrhotic lesions of the liver parenchyma or not (see Chapter 8).

II. Evaluation of the intensity of the obstruction

Portal pressure has always been considered the basic element of the hemodynamic classification of portal hypertension. Viamonte proposed a three-stage classification based on the corrected sinusoidal pressure: mild portal hypertension, from 6 to 14 mm Hg; moderate, from 15 to 30 mm Hg, and severe, above 30 mm Hg (22–23).

This interesting classification is insufficient because portal hypertension is not only a consequence of an obstruction to the portal flow but also dependent on the development of a collateral network. For this reason, Reuter has classified the post-sinusoidal obstructions as a function of the decrease of the hepatopetal portal flow (15). This decrease in hepatopetal portal flow correlates well with the intensity of the obstruction and therefore with the severity of the primary disease.

By injecting contrast medium of low viscosity (60 ml in 10 seconds) in the superior mesenteric artery, the venous phase is almost always of high quality. Arterial portography evaluates the decrease in hepatopetal portal flow directly by the poor quality of opacification of intrahepatic portal branches and portal hepatography, and indirectly by two signs: the development of a hepatofugal porto-caval circulation and the hepatic arterial circulation.

At the venous phase of superior mesenteric arteriography, venous flow can be considered as proportional to the diameter of the vessels and to the density and the extent of opacifition in the corresponding territory.

In physiologic conditions, with a normal hepatopetal portal flow, the portal trunk and the intrahepatic portal branches are clearly opacified, producing a good portal hepatography at the 16th second. Under pathologic conditions the relative values of the hepatopetal and hepatofugal portal flows can be evaluated by the diameter of the vessels and the quality of opacification in the diverse territories.

The dilatation of the hepatic artery and especially that of its segmental intrahepatic branches and the increase in arterial hepatography is also proportional to the decrease in hepatopetal portal flow except in the Budd Chiari syndrome, with its particular hemodynamic changes and its characteristic angiographic pattern. It is then possible for the post-sinusoidal portal hypertensions due to intrahepatic obstruction to be classified according to the degree of the obstruction. Four stages can be distinguished.

1. Minimal obstructions

Minimal obstructions are responsible for little or no decrease in hepatopetal portal flow. Thus, during the superior mesenteric venous phase the intrahepatic portal branches are well opacified and sometimes the portal hepatography persists at the 15th second. However, at this stage, the decrease in portal flow can be detected by the persistence of the opacification of the intrahepatic portal branches which normally are no longer visible. At the same time, the hepatofugal porto caval derivations are not opacified and the caliber of the hepatic artery is normal.

Under these conditions, the portal hypertension is minimal, and usually clinically latent. The angiographic diagnosis is based on the splenic pattern which varies according to both the portal pressure and the duration and progression rate of the obstruction.

If the portal pressure is only slightly elevated or even subnormal, the spleen can be normal. This situation can only arise as a consequence of spontaneous functional pre-existing portocaval anastomoses which must be visible on the venous phase of the superior mesenteric arteriography. It is possible that the spleen remains normal in these conditions, even with a tight obstruction to the hepatopetal portal flow. This is even more likely if the obstruction is minimal, but this is quite rare in practice (Fig. 33).

If the obstruction is recent, a moderate splenomegaly with a slightly dilated and elongated splenic artery can be observed. The intrasplenic circulation is more or less normal and the splenic vein is opacified correctly. The dilution of the contrast medium is the only sign of a moderate splenic stasis (Fig. 5).

In case of long-standing and stationary obstruction a voluminous splenomegaly can be observed. The splenic artery is very dilated as well as the splenic vein, the opacification of which is all the more weakened as the spleen is more enlarged. This is the angiographic pattern found most frequently. The moderate and slowly progressive obstructions remain clinically latent for many years until they are discovered by finding a voluminous splenomegaly often associated with signs of hypersplenism (Figs. 15, 17, 59, 60, 61, 62, 63).

In our experience, these voluminous splenomegalies seem to be the constant result of an old minimal and slowly progressive obstruction, regardless of the etiology. Two essential conditions seem to be required for the development of a stasis related voluminous splenomegaly. The venous stasis must be minimal; it must not slow down the intrasplenic circulation too much: and it must be of long duration: the voluminous splenomegaly probably needs years to develop.

2. Moderate obstructions

The decrease in the hepatopetal portal flow can be seen by the absence of portal hepatography on the late films of the superior mesenteric venous return and by the poor opacification of the distal intrahepatic portal branches. Only the segmental portal branches and a few of their collaterals are well opacified. This decrease of the hepatopetal portal flow inside the liver is usually diffuse and homogenous (Fig. 46) but it can also be nonhomogenous and irregularly distributed in the various hepatic segments. Some segmental portal branches are more opacified than others, and the late films demonstrate a nonhomogenous hepatography (Fig. 47). At the same time, the hepatofugal collateral circulation most often appears as a moderate reflux in the inferior mesenteric and left gastric veins whose diameter remains normal or only slightly dilated (Fig. 46).

At this stage with a moderate hepatofugal flow, the opacification of the porto-caval derivations vary especially as a function of each patient's anatomical potential, and it seems obvious that the portal hypertension will be greater as these collaterals are less developed. It is thus posssible that the retrograde opacification of the left gastric vein is not visible on arteriography even if there exists voluminous esophageal varices if the hepatofugal flow of these varices remains low. In these conditions, an important dilation of the intestinal veins, which are tributaries of the superior mesenteric veins, indicates a venous stasis which involves not only the splenic territory but also the mesenteric one (Fig. 34). However, the decrease in hepatopetal portal flow is confirmed by the moderate dilation of the hepatic artery, which can be well visu-

alized at the intrahepatic branch level.

In these cases the splenomegaly is quite variable depending on the duration of the disease and the level of the portal hypertension, the latter partially depending on the development of a collateral hepatofugal circulation. If the disease is of long duration and the collateral circulation well developed, the splenomegaly can be quite voluminous; if the collateral circulation is poorly developed with a greater portal hypertension, the splenic circulation is slowed due to the stasis and the splenomegaly remains moderate.

3. Severe obstructions

If the intrahepatic obstruction is more severe, the hepatopetal portal flow is greatly diminished: the portal vein remains well opacified but the intrahepatic portal branches are quite thinned out, only the large intrahepatic portal branches are opacified and their diameter is decreased.

Otherwise the collateral hepatofugal circulation is well visible with retrograde opacification of the inferior mesenteric vein, the dilated left gastric vein, and sometimes even the splenic vein. The diameter of the left gastric vein approximates that of the portal vein which is decreased. On the whole, the collateral circulation is better opacified than the hepatic territory; this suggests that the hepatofugal circulation dominates the hepatopetal circulation (Fig. 48).

The hepatic artery and its intrahepatic branches are quite dilated and the arterial hepatography is dense. Under these conditions, the selective hepatic arteriography can demonstrate a retrograde opacification in one or more intrahepatic portal branches. This corresponds to the first stage of the inversion of the intrahepatic portal flow. In these circumstances the splenomegaly is usually minimal or moderate because the intrasplenic circulation is greatly reduced (Figs. 4, 40 48).

4. Very severe obstructions

The complete interruption of the hepatopetal portal flow is demonstrated by the absence of intrahepatic portal branch opacification. The portal vein itself may either remain visible to the level of the hilus or not be opacified at all. Therefore, the portal flow is completely hepatofugal. This is demonstrated by the presence of voluminous varices opacified via the very dilated gastric veins and often by the spontaneous and functional portocaval anastomoses. The opacification of these anastomoses can sometimes be followed all the way to the inferior vena cava.

The liver is vascularized only by the hepatic artery which is extremely dilated, and the retrograde opacification of the intrahepatic portal branches is frequent following the dense arterial hepatography. In certain cases the obstruction is so severe that most of the arterial blood drains in a retrograde manner into the portal vein which is totally hepatofugal: the inversion of the portal blood flow is complete (Figs. 42, 43, 44, 45).

This type of obstruction usually corresponds to rapidly progressive lesions which precociously interrupt the splenic blood flow. Generally, the splenomegaly is quite minimal. However, owing to a high flow rate porto-caval anastomosis a severe obstruction may be associated with a minimal portal hypertension. As a result of this the angiographic spleen pattern may be normal (Fig. 49).

III. Etiological diagnosis

Post-sinusoidal obstructions are caused by different processes. Their etiological diagnosis is often quite difficult on the basis of clinical, or even histological criteria.

Alcoholic cirrhosis is the most frequent cause, and this diagnosis is evoked from the start, sometimes incorrectly in patients who may be considered alcoholics. On the other hand, arterioportography is frequently performed after discovery of a hepatic anomaly or signs of portal hypertension (esophageal varices, splenomegaly or ascites) without knowing the exact nature of the illness. Arterioportography can contribute to the etiological diagnosis by demonstrating the location of the obstruction and sometimes even its nature.

Concerning the location of the obstruction, it is relatively easy to distinguish between the intrahepatic obstructions and the suprahepatic obstructions whose hemodynamic consequences are dif-

ferent. In the first case, the obstruction is progressive and usually incomplete. It is associated with diffuse hepatic lesions. In the second case, however, the obstruction is most often complete and appears rapidly on an otherwise normal liver; the modifications observed on arterioportography are therefore completely different.

1. Suprahepatic obstructions

Suprahepatic obstructions are caused by obstruction of the large hepatic veins (Budd-Chiari syndrome) and more rarely by an obstruction of the small intrahepatic veins (veino-occlusive disease) or by cardiac lesions responsible for an inferior caval hypertension (constrictive pericarditis).

Hemodynamically, Budd-Chiari syndromes are differentiated according to whether or not the obstruction of the hepatic veins is associated with the obstruction of the inferior vena cava. When it is associated, the Budd-Chiari syndrome is secondary either to a hepatic tumor or to a retroperitoneal tumor (cancer of the right kidney, for example), or to a web of the inferior vena cava presumably of congenital origin.

The obstruction of the hepatic veins can be isolated and due to a thrombosis or associated with a myelo-proliferative disease or the taking of oral contraceptives, or of unknown etiology. These Budd-Chiari syndromes with isolated thrombosis of the hepatic veins, sometimes called primitive Budd-Chiari syndrome, seem to be the most frequent. Of the 15 cases we have explored, 12 correspond to an isolated thrombosis of the hepatic veins and 3 to a congenital obstruction of the inferior vena cava. The Budd-Chiari syndromes which are secondary to tumors are less interesting diagnostically because the signs of the responsible tumor usually predominate.

Arterioportography is quite interesting in the diagnosis of the isolated thrombosis of the hepatic veins because this diagnosis is often difficult using clinical and biological information.

The methods of morphologic exploration, e.g., scintiphotography and ultrasound, can be misleading. However, the discovery on ultrasonography of a large sonolucent caudate lobe is at the present time a good sign of orientation. Biopsy can show signs of congestion and of centro-lobular necrosis which can evoke the diagnosis. However, it is usually confirmed only by hepatic phlebography, which shows the characteristic spider's web pattern of the collateral venous network.

Certain authors have stressed the particular angiographic anomalies, such as a thinning and a stretching of the intrahepatic arterial ramifications (47, 50, 58, 59) or an inversion of the portal flow (47). However, the diagnostic reliability of arteriography in this syndrome has not been stressed enough.

The angiographic pattern is quite characteristic and constant in all cases because of the special circulatory changes which are the consequence of the isolated thrombosis of the hepatic veins. Because the obstruction of the hepatic veins is usually rapidly progressive, the sinusoidal pressure increases dramatically so that not only the hepatopetal portal flow is completely interrupted but the hepatic arterial flow is also decreased and the intrahepatic arterial ramifications remain abnormally thin. This angiographic pattern corresponds hemodynamically to the syndrome of hepatic ischemia of venous origin. It is identical to what can be observed everywhere else in the organism when a significant obstruction of an efferent vein leads to ischemia in the corresponding territory. For example, this can be observed in case of intestinal ischemia (27, 28) as well as in some cases of lower limbs phlebitis (phlegmasia caerulea).

The angiographic pattern of the Budd-Chiari syndrome is quite characteristic. To our knowledge, it is the only circumstance where a complete, or almost complete, obstruction of the hepatopetal portal flow is not associated with a compensatory increase in the hepatic arterial flow. The following signs in arterio-portography are typical of this diagnosis:

a) The portal vein is usually patent, although portal thrombosis may be associated to the hepatic vein thrombosis in approximately 20% of the cases. In such cases (2/12 cases in our experience), the evaluation of the decrease of the hepatopetal portal flow is of course impossible (Fig. 25).

In most cases, however, the poor opacification of the intrahepatic portal branches corresponds to a severe or very severe obstruction, while the hepa-

tofugal flow is usually poorly developed. The contrast medium stagnates in the dilated intestinal or gastric veins because the arterioportography is usually performed at an early stage of the disease.

The obstruction to hepatopetal portal flow is either diffuse throughout the hepatic territory when all the hepatic veins are obstructed or predominates in certain territories while a certain portal flow persists in the other territories, which sometimes leads to the persistence of a portal hepatography in certain segments (Fig. 55). The caudate lobe drains directly into the inferior vena cava by small accessory hepatic veins which usually remain patent, explaining the compensatory hypertrophy of this segment. In fact, it is normal that the portal flow persists where the hepatic drainage persists and when the hepatic venous thrombosis is partial or incomplete, we have always obtained an excellent correlation between the findings of hepatic phlebography and portography.

b) Hepatic arteriography demonstrates a diffuse hepatomegaly which often predominates on the left lobe, sometimes with a pseudo-tumoral aspect, with a downward displacement of the hepatic artery trunk, when an important hypertrophy of the caudate lobe exists. It is essential to note that the hepatic artery is not dilated and the intrahepatic arterial ramifications are on the contrary elongated, stretched and regularly spread apart from each other. The regular passive distension of the hepatic parenchyma is due to the diffuse dilation of the sinusoidal capillaries secondary to blood stasis (Figs. 10, 50, 51).

The intrahepatic arterial ramifications are thin and the arterial hepatography is quite weak. There is no increase in arterial flow to compensate the decrease in portal flow.

Because the obstruction of the hepatic veins is not always complete and that a collateral portohepatic circulation can develop mainly from the sub capsular portal venous network, some hepatic territories may remain better perfused than others. This explains the nonhomogenous pattern of the arterial hepatography, with some dense nodular opacities. These have been reported to simulate metastatic nodules (60) (Fig. 51).

In two cases we have been able to observe a very particular hepatographic pattern characterized by the opacification of the outline of the liver by a thin stripe of contrast medium. This may be due to passage of the contrast medium injected arterially into the subcapsular venous portal network. This may confirm the existence of subcapsular arterial-portal anastomoses described by Chenderovitch using micro-angioradiography (1) (Fig. 52).

Arterio-portal communications can bring about an inversion of the intrahepatic portal flow and can be observed in the Budd-Chiari syndrome, as well as in all other severe post-sinusoidal obstructions regardless of their cause or location. However, the inversion of the portal flow is much more discrete and much less apparent in Budd-Chiari syndrome than in cirrhosis as it depends equally on three factors: 1) the increase of hepatic arterial flow; 2) the development of intrahepatic arterio portal communications; and 3) the hepatofugal portal flow.

Cirrhosis perfectly fulfills all three of these conditions, while on the contrary, during the Budd-Chiari syndrome the arterial flow is not increased, there are no abnormal intra-hepatic arterio-portal communications, and the hepatofugal portal flow is often not very developed. Thus this sign is quite inconsistent (7/12 cases) and discrete, especially at the beginning of the evolution, and limited to the retrograde opacification of a few intrahepatic portal branches. If the portal trunk is opacified, its opacification is quite weak and may remain unknown (Figs. 53 & 54).

If the patient survives, which depends on the possibilities of the development of the collateral circulation, the hepatic arterial flow increases progressively, as does the hepatofugal portal flow. The inversion of the intrahepatic portal flow then becomes evident (Fig. 55).

c.) The splenic pattern is just as peculiar: because the supra-hepatic obstruction is quite severe and rapidly constituted, the thrombosis of the hepatic veins is complete in a few days or weeks; the rapid increase of portal pressure leads to an almost complete interruption of the splenic circulation. Thus, the spleen is only slightly hypertrophied, or not at all, and the splenic artery, especially the intrasplenic ramifications, is particularly thin. This corresponds to the low flow rate pattern of the spleen, which finds its most characteristic example in the

Budd-Chiari syndrome (Figs. 10, 50, 53, 55).

Of course, the splenomegaly is more minimal as the evolution is more recent and more acute. On the contrary, if the patient survives after, for example, a surgical portocaval anastomosis, which allows the venous portal blood to drain again, a very large splenomegaly will develop after a few years (Fig. 10).

Finally, because of the large decrease of the hepatic arterial flow and of the splenic arterial flow, the celiac arteriography will also often demonstrate abnormally dense opacification of the gastric and pancreatic territories (Fig. 50).

Similar angiographic modifications have been observed in the 12 cases of isolated obstructions of the hepatic veins confirmed by hepatic phlebography. We feel that arterio-portography is a very reliable method for the diagnosis of the Budd-Chiari syndrome.

These anomalies of intrahepatic circulation during the Budd-Chiari syndrome by isolated thrombosis of the hepatic veins are sometimes well demonstrated by computed tomography with vascular injection (61, 62). We have observed this in four cases.

The inversion of the intrahepatic portal current is shown by the early opacification of the intrahepatic portal branches at the arterial stage, that is, on the first section 10 seconds after the rapid intravenous injection of the contrast medium (Fig. 54). On the contrary, 10 to 20 seconds later, at a stage where normally the intrahepatic portal branches are opacified, these portal veins are no longer visible and there is no portal hepatography.

This angio-C.T. pattern of the inversion of the intrahepatic portal flow is particularly obvious when this inversion of the portal flow is localized, or at least predominates in certain segments. This is frequent in the Budd- Chiari syndrome. The early opacification of certain intra-hepatic portal branches contrasts with the absence of visibility of the portal veins in the other territories. Of course, the arteriography confirms this hemodynamic anomaly by demonstrating a perfect correlation between the early film of the angioscan and the hepatic arteriography film 4 to 7 seconds after injection (Fig. 54).

The computed tomography sections taken 20 to 40 seconds after injection and corresponding to the stage of portal opacification are the most interesting ones. On these sections, we can evaluate the modifications of the intrahepatic hepatopetal portal flow more precisely than on the venous phase of the superior mesenteric arteriography.

Sometimes the portal hepatography is absent or homogeneously weak, which corresponds to a complete obstruction of all the hepatic veins. More often, the portal hepatography persists at the hypertrophied caudate lobe, while it is lacking in the other territories (Fig. 54). In other cases, some segments of the left lobe remain opacified as well, while the portal hepatography of the right lobe is completely absent. This indicates that the obstruction of the hepatic venous system is complete on the right and partial on the left, which can be confirmed by hepatic venography (Fig. 53).

In other cases, the hepatography is completely nonhomogeneous, which corresponds to a complete obstruction of the hepatic veins with an interruption of the hepatopetal portal flow whereas collateral circulation persists in some territories giving rise to a stagnation of the contrast medium. This pattern probably has the same significance as the nonhomogeneous arterial hepatography visible on the late films of the celiac arteriography (Figs. 56 & 57). Finally, the hepatic veins are never visible on the sections passing by the superior part of the liver.

In the four observations where a computed tomography was performed, there was an excellent correlation with the angiographic explorations. We feel that computed tomography is quite useful for the diagnosis of the Budd-Chiari syndrome because it details precisely both the morphologic and hemodynamic hepatic modifications.

When the obstruction of the hepatic veins is associated with an obstruction of the inferior vena cava, e.g., in congenital Budd-Chiari syndrome, we have observed the same arteriographic modifications of the liver and of the spleen as in the preceding cases. However, the obstruction of the hepatic veins is by definition complete and the intrahepatic circulation depends essentially on the development of the collateral circulation between the inferior vena cava and the superior vena cava. The hepatopetal portal flow is completely blocked, including the caudate lobe. On the computed tomography sections the hepatography can be either

homogeneous or slightly nonhomogeneous, but the diagnosis can be suggested by the abnormal opacification of the perirachidial veins which corresponds to a collateral circulation between the inferior and superior vena cava systems.

The cardiac lesions responsible for an inferior vena caval hypertension create a functional obstacle to the hepatic efferent flow. The resulting hemodynamics are similar to those of congenital obstructions of the inferior vena cava.

In one case of constrictive pericarditis we have been able to verify that the angiographic modifications are identical to those observed in the Budd-Chiari syndromes. The pattern of the liver in particular is characterized by a diffuse hepatomegaly. The intrahepatic arterial ramifications are thin, taut, elongated, and regularly spread out. The fact that these two patterns are identical proves that they are a consequence of the passive distension of the normal hepatic parenchyma secondary to the stasis in the sinusoid capillaries (Fig. 58). The diagnosis is easily confirmed by the hepatic venography, which demonstrates that the hepatic veins are not obstructed but dilated.

2. *Intrahepatic obstructions*

Contrary to the Budd-Chiari syndrome whose angiographic pattern is quite characteristic, intrahepatic obstructions arise from multiple causes and do not present a particular angiographic pattern. However, on arterio-portography we can recognize that the obstruction is intrahepatic when the angiographic pattern is not that of the Budd-Chiari syndrome, and the portal vein is patent. It is of little use for the etiological diagnosis which depends essentially on the histological study of the hepatic parenchyma.

It must be understood, however, that precise etiological diagnosis of the intrahepatic obstruction is often quite difficult on the basis of the clinical, biological, and even the histological data. Liver biopsy is sometimes insufficient to lead to a precise diagnosis (63).

Of course, alcoholic cirrhosis is the most frequent cause and it is usually clinically evident by the association of the signs of portal hypertension, hepatic insufficiency, and alcoholic intoxication. It is probable that alcoholic cirrhosis is overdiagnosed. A systematic histological study of the liver in alcoholic patients has demonstrated that some hepatic lesions have a different etiology (64). In fact, numerous hepatic lesions, some of which are poorly known, can be revealed by signs of portal hypertension which often evolve slowly without serious repercussions on the hepatic cellular function. For this reason, and because of the discretion of certain histological lesions of the liver, portal hypertension has sometimes been considered as idiopathic.

At the present time, these non-cirrhotic hepatic lesions seem more frequent. Some are better known, but the diagnosis remains quite difficult. For these reasons the angiographic patterns should be discussed, even if they are of limited interest. They are useful in the orientation of the diagnosis and for the physiopathology.

From a practical point of view, these lesions can be classified in three groups: (a) alcoholic cirrhosis and post-necrotic cirrhosis, (b) other well-individualized hepatopathies, and (c) peri-portal fibrosis (idiopathic non-cirrhotic portal hypertension). Some of these lesions do not correspond to a post-sinusoidal obstruction, but rather to a pre-sinusoidal or mixed obstruction. However, there is no correlation between the angiographic patterns and the intrahepatic site of the obstruction, which can be demonstrated only by *hepatic venography* with the measurement of pressures.

a) Alcoholic and post-necrotic cirrhoses

There is no specific angiographic sign of these diverse causes of intrahepatic obstruction, or of the cirrhoses in particular. The angiographic pattern is principally a function of the degree and the evolving nature of the obstruction.

However, during the cirrhoses, the liver rarely maintains a normal morphology; it is either hypertrophied, atrophied, or atropho-hypertrophied. The intrahepatic arteries are more or less tortuous depending on the degree of liver atrophy and the peripheral arterial network is often well developed, the distal ramifications are usually numerous and sometimes seem duplicated.

However, in most of these cases, the angiographic patterns of these alcoholic cirrhoses and those of post-hepatitis cirrhoses differ on two

points: splenic volume and the pattern of the liver and the hepatic artery.

The splenomegaly is in fact of more consequence during post-necrotic cirrhoses than during alcoholic cirrhoses. This was originally reported by Abeatici and confirmed by our experience. In 280 cases of alcoholic cirrhosis, the long axis of the spleen averages 16 cm, while in a group of 26 post-hepatitis cirrhoses, it averages 19 cm (65). Of course, this very large splenomegaly is accompanied by a dilation of the splenic vessels and particularly of the splenic vein and the portal vein, while the splenic vein is often normal or dilated very little in alcoholic cirrhoses. Several observations of post-hepatitis cirrhoses which were discovered late because of an apparently primitive splenomegaly associated to a hypersplenism have been reported in the literature (66, 67, 68, 69). The hypersplenism has been considered a frequent sequela of epidemic hepatitis.

This more serious splenomegaly signifies that the evolution of the obstructive intra-hepatic lesions is slower and usually less severe during post-hepatitis cirrhoses than during alcoholic cirrhoses. This agrees with various authors who have noted that the functional liver alteration is quite moderate, and that the cirrhosis occurs quite late after the initial episode of hepatitis (70).

In terms of the liver, post-hepatitis cirrhoses are also distinguished from alcoholic cirrhoses by the fact that the hepatic artery is slightly dilated or not at all, and that the liver is severely atrophied.

The dilation of the hepatic artery is quite constant and often serious during alcoholic cirrhoses, while in many post-hepatitis cirrhoses the caliber of the hepatic artery remains within normal values and can sometimes be thinner than normal.

Liver atrophy is also an interesting sign of post-hepatitis cirrhoses, but when it is diffuse, this atrophy can also be observed in alcoholic cirrhoses. In this case, however, the diffuse atrophy of the liver is observed especially at a late stage of the disease and is accompanied by a serious dilation of the hepatic artery, which is not the usual case during post-hepatitis cirrhoses. However, the partial or segmental atrophy seems more characteristic of post-hepatitis cirrhoses. In this disease, an extremely serious atrophy of certain lobes or hepatic segments can be observed, while other parts of the liver are normal or, on the contrary, hypertrophied (Figs. 59, 63). Contrary to alcoholic cirrhosis, this liver atrophy is not always accompanied by portal hypertension, or else this portal hypertension is moderate, clinically latent, and remains unknown. Thus, it is often a source of error of interpretation. For example, when the atrophy of the right lobe is accompanied by an upward and outward displacement of the gallbladder, which is habitual, this abnormal location (aberrant gall bladder) can be wrongly considered as a congenital anomaly associated to a hypoplasia of the right lobe of the liver.

In summary, these angiographic differences seem to correspond principally to a different progression of the intrahepatic obstruction: the alcoholic cirrhoses correspond more often to a tight intrahepatic obstruction which evolves rapidly, while post-hepatitis cirrhoses frequently present the pattern of a minimal or moderate intra-hepatic obstruction which evolves slowly (Figs. 11, 60, 61).

However, it is possible that the anatomic lesions of post-hepatitis cirrhosis are responsible for the atrophic alterations and the limited development of the arterial circulation.

These angiographic differences are, of course, inconsistent, and their practical interest is debatable. In the large series we have observed, however, they are frequent enough to merit attention.

b) Other well-individualized hepatopathies

Numerous hepatic diseases can lead to portal hypertension during their evolution. The portal hypertension is often moderate and late, and does not create diagnostic problems because the responsible illness is easily identified by its own symptomatology.

1. Hemochromatosis and Wilson's disease can lead at a late stage to a cirrhosis whose progress is usually slow and benign. The portal hypertension is late and often well tolerated. Its primary sign is a splenomegaly which is often very large (Fig. 13).

2. Pure hepatic steatosis has also been described as a possible cause of portal hypertension by an intrahepatic block, whether it is pre- or post-sinusoidal (71, 72, 73, 74).

3. Sarcoidosis is also a rare cause of portal hypertension whose mechanism is not known (73, 75).

4. Chronic hepatitis following viral hepatitis and

chronic active hepatitis are also accompanied by portal hypertension. Certain authors have demonstrated that a moderate, latent portal hypertension appears frequently at an early stage of post-viral chronic hepatitis (76, 77).

On angiography, these are indicated only by a discrete splenomegaly; the increase in volume can be seen in successive examinations.

5. Acute viral hepatitis is frequently accompanied by portal hypertension, which is usually regressive in the patients who survive (78).

6. Acute alcoholic hepatitis is equally a classic cause of portal hypertension.

7. Some hepatic tumors, diffuse hepatic metastases in particular, can also lead to an obstruction of the hepatopetal portal flow.

In all these cases the portal hypertension is usually moderate and secondary to the signs of the responsible illness. Angiography usually indicates a pattern of minimal or moderate portal hypertension which evolves slowly. The hepatic artery is slightly dilated, or not at all, a hepatofugal collateral circulation which is not well-developed, a more or less large splenomegaly which corresponds to the duration of the disease, and a diffuse hepatomegaly.

8. Partial nodular transformation of the liver is also a rare disease which is accompanied by portal hypertension in 50% of the cases. The diagnosis is difficult and depends on the biopsy, which demonstrates nodular transformation. These nodules are not surrounded by disseminated fibrosis but are found irregularly in the hepatic parenchyma. This disease is often latent and is not accompanied by signs of hepatocellular insufficiency. It can be associated to another disease such as Felty's syndrome or myeloid selenomegaly (63, 79, 80).

c) Idiopathic non-cirrhotic portal hypertension

This group of diseases corresponds to cases where the arterioportography suggests an intrahepatic obstruction which may correspond to a cirrhosis. Neither the diagnosis of cirrhosis nor that of other known hepatic diseases can be confirmed by the clinical and biological data and the hepatic biopsy. These cases have been described by different authors in different countries under a variety of names: idiopathic portal hypertension (81), hepatoportal sclerosis (82), essential portal hypertension (83), portal or periportal fibrosis (84, 85), Banti's disease, cirrhosis of splenic origin (86), and Mediterranean cirrhosis (87).

At first, these diseases were described mainly in the Mediterranean region. They were revealed by a very large splenomegaly associated to hypersplenism and later by digestive hemorrhaging. However, the liver seemed functionally and histologically normal. For these reasons, certain authors thought that it was a 'forward pressure' portal hypertension of splenic origin.

However, after a relatively long evolution, Banti's disease always leads to a hepatic insufficiency associated to fibrotic lesions or even a true hepatic cirrhosis. Some authors have thought that these hepatic lesions were a consequence of the splenomegaly, even though splenectomy has never prevented the evolution of the disease and the aggravation of the hepatic lesion (Fig. 62).

On the contrary, because of our experience, we think that the very large splenomegaly is only the late consequence of a minimal intrahepatic obstruction which progresses slowly and which can easily escape the hepatic biopsy

Different authors have noted that these portal hypertensions of unexplained origin without histological signs of cirrhosis are accompanied by lesions of hepatic fibrosis. These fibrotic lesions are usually moderate and have a periportal location. Their role as an etiology of portal hypertension has been questioned. This is why these cases have been reported under a variety of names.

Some authors have felt that these lesions were too discrete to have hemodynamic significance and that the liver was normal or almost normal histologically. They concluded that the portal hypertension was without obstruction and identified these cases as 'essential portal hypertension' or 'portal hypertension of splenic origin.'

On the contrary, other authors using the terms 'idiopathic portal hypertension' or 'non-cirrhotic portal fibrosis' considered the hepatic lesions to be responsible for the portal hypertension in determining a pre-sinusoidal obstruction. This obstruction has been demonstrated either by angiographic methods, i.e., trans-hepatic portography, which can demonstrate a pattern of distortion and amputation of the small intra-hepatic portal branches,

or by anatomical studies which have shown after post-mortem injection of the liver anomalies which evoke microthrombosis of the small intrahepatic portal veins (81, 88, 89, 90).

In our experience, these histological lesions of periportal fibrosis are frequently found at hepatic biopsy in non-alcoholic patients who present a portal hypertension whith a very large splenomegaly and few signs of functional hepatic alteration. In these patients there are frequently serious differences between the angiographic and macroscopic patterns of the liver and the histological lesions demonstrated by hepatic biopsy. In some cases the periportal fibrotic lesions are very discrete, and the histological pattern of the liver seems almost normal, while arteriography demonstrates a serious hepatic atrophy, whether diffuse or segmental, and during surgical exploration an additional micro- or macronodular pattern can be observed. On the contrary, in other cases the liver can appear normal or only slightly, modified macroscopically, while the biopsy demonstrates serious lesions of periportal fibrosis.

As the atrophic patterns of the liver are particularly frequent in these patients, these angiographic signs which demonstrate the existence of serious hepatic lesions must be emphasized, while the liver can appear histologically normal, or almost normal. This discrepancy can perhaps be explained by the hepatic transformation which is often atropho-hypertrophic. The biopsy has a greater chance to select the hypertrophic rather than the atrophic segments which are more deeply situated in the liver. The histological lesions of the hypertrophic segments are likely to be less severe than on the atrophic segments (Fig. 63).

In an earlier study (65) including these cases of periportal fibrosis as well as a few observations of cirrhosis of unknown origin, we noted that patients can be classified into three different groups according to their clinical context. The first group corresponds to the diseases of Mediterranean origin, especially North African, whose clinical picture is similar to that of Banti's disease. The second group corresponds to patients who have presented in their antecedents an episode of viral hepatitis so that we are led to believe that these fibrotic lesions or lesions of hepatic cirrhosis are a long-term complication of the earlier hepatitis. In a third group of cases no explanation can be found for the origin of the portal hypertension which is apparently idiopathic.

In these three groups of patients arterioportography demonstrates the same patterns of minimal or moderate portal hypertension with a very large splenomegaly. The liver is often atrophic and the hepatic artery is either slightly dilated or, on the contrary, thinner than usual. These patterns resemble those which are observed in most cases of post-hepatitis cirrhosis. In addition, in about 30% of the patients in the first and third groups we find anatomical anomalies (macronodular pattern of the liver) or biological anomalies (Hb As +) which suggest also that these lesions could be of viral origin. These results suggest that it is possible in certain cases, apparently idiopathic portal hypertensions are in fact sequelae of an unknown may be anicteric viral hepatitis which occurred previously.

The frequency of anicteric forms of viral hepatitis has been stressed by numerous authors, as well as the delayed onset of the cirrhogenous evolution (76). Finally, the frequency of viral hepatitis in Mediterranean countries is well known, and certain authors, studying the histological pattern of the liver in some of these subjects, healthy carriers of Australia antigen, have demonstrated that there is often a periportal fibrosis which they consider as the probable sequela of the previous viral disease (91).

Finally, a great number of medications can cause a toxic hepatitis which may be capable of evolving toward chronicity. The products incriminated most often are: oxyphenisatin, alphamethyldopa, nitrofurantoin, and chlometacin (92). Thus it is probable that certain drug intoxications are a frequent cause of chronic hepatitis and certain authors believe that 20 to 40% of chronic hepatitis with negative Australia antigen are in fact caused by drugs (93).

A few cases of periportal fibrosis secondary to arsenic intoxication or exposure to vinyl chloride have been reported by authors who suggest that idiopathic portal hypertension could be related to domestic or industrial exposure to hepatotoxins (90)

Idiopathic portal hypertension after renal transplantation probably due to hepatotoxicity of immunosuppressive drugs was also reported. This would

create a perisinusoidal fibrosis which would be invisible under light microscopy but always present in electron microscopy. This could be the very early stage of hepatoportal sclerosis (84, 85).

Finally, the partial nodular transformation of the liver is often associated to a periportal fibrosis and its angiographic pattern is identical to that of idiopathic portal hypertension. In particular the hepatic artery is often thinner than normal (80, 88). Therefore, according to Shedlofsky, it is possible that these entities correspond to two anatomical expressions of the same pathological process.

Even though this problem is still poorly understood, it seems that most cases of idiopathic portal hypertension reported in different countries under various names include numerous similarities clinically, angiographically, and anatomically. It is probable that they belong to the same group of diseases.

The typical clinical pattern is that of a moderate portal hypertension which evolves slowly and is revealed either by a splenomegaly or by digestive hemorrhaging in relation to esophageal varices. It is always accompanied by a very large splenomegaly, sometimes associated to a hypersplenism. The liver is often atrophic or atropho-hypertrophic and the hepatic artery is slightly dilated or sometimes even thinner than normal.

The functional hepatic tests remain normal or slightly disturbed. The increase of portal pressure is moderate, usually inferior to 20 mm of mercury. The study of hepatic vein pressures shows that the obstruction can be either pre- or post-sinusoidal, or both pre- and post-sinusoidal.

Anatomically, the surface of the liver can be either macro- or micronodular, as in cirrhosis, or can appear almost normal. Histologically, the only sign can be a periportal fibrosis. when the histological lesions are discrete, they may be visible only on a surgical biopsy, the needle biopsy often being considered normal or nearly normal. If in these conditions the right lobe of the liver is very atrophied, which is not rare in our experience, it is understandable that with the normal or nearly normal results of the functional hepatic tests and of the hepatic biopsy the portal hypertension is incorrectly considered to be due to a congenital hypoplasia of the right lobe of the liver. In fact we believe that it is unlikely that congenital hypoplasia of one hepatic lobe could lead to a portal hypertension due to the regenerative capacity of the other lobe. In addition a right hepatectomy in adults does not lead to portal hypertension.

On the contrary we have frequently observed this segmental atrophy of the liver during idiopathic portal fibrosis and hemodynamically it is probable that a portal fibrosis, even if it is very discrete, is capable of leading to a minimal portal hypertension because the portal system is one of high flow and weak pressure gradient.

Until we understand more thoroughly the physiopathology and the etiologies of the acquired idiopathic periportal fibrosis, they should be grouped according to their angiographic pattern, which seems interesting diagnostically and physiopathologically.

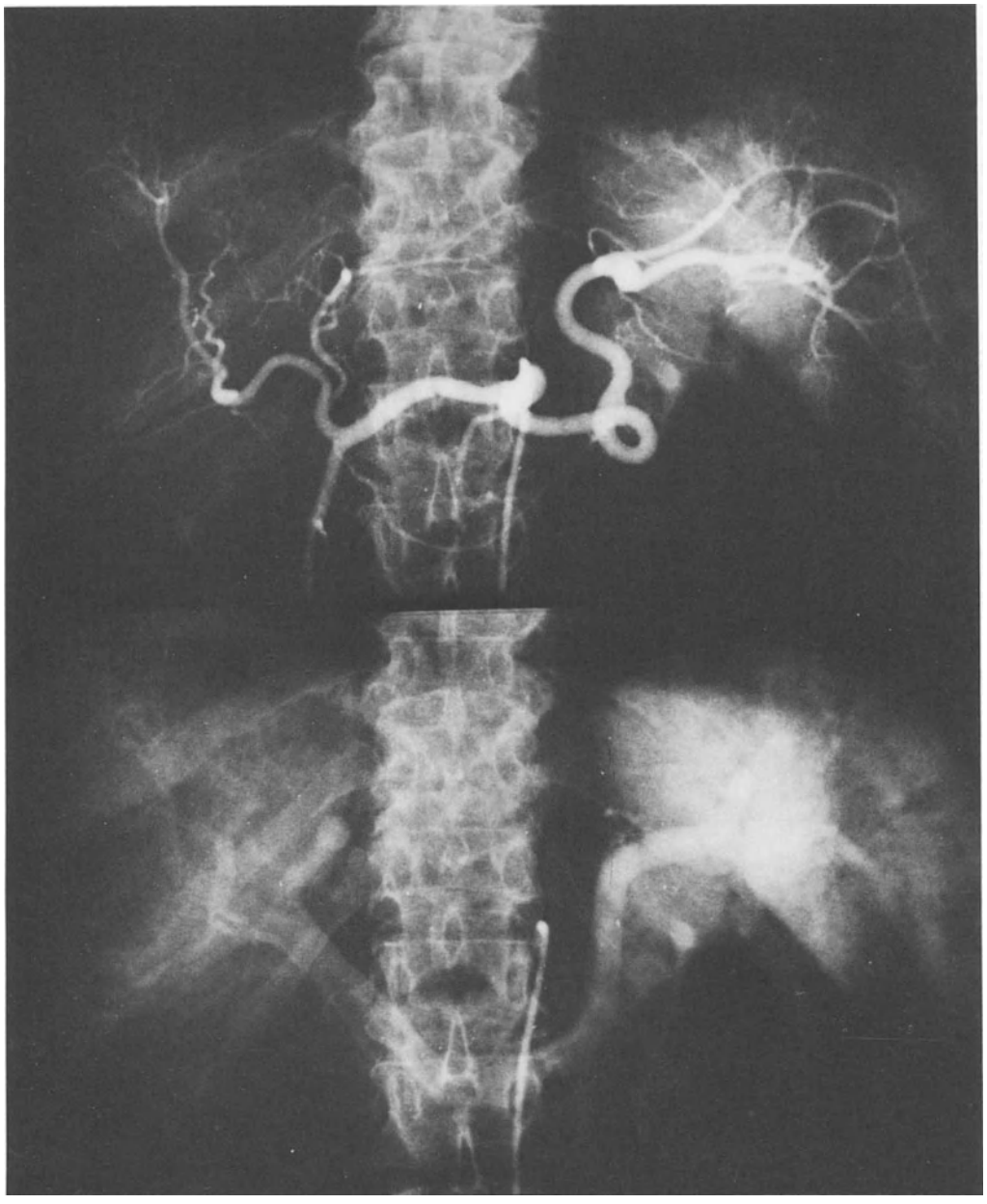
Diagnostically, the angiographic pattern sometimes suggests a cirrhosis and it is the biopsy which demonstrates that it is in fact a non-cirrhotic periportal fibrosis.

However, in a number of cases arterioportography demonstrates a pattern of minimal or moderate portal hypertension by a longstanding intrahepatic obstruction which evolves slowly with a very large splenomegaly and a liver which is either normal or, more often, atrophied. The dilation of the hepatic artery could be absent because the hepatopetal portal flow is only slightly decreased; it could also be in part a consequence of the particular anatomic lesions of this periportal fibrosis. This would explain why the intrahepatic arterial ramifications are sometimes abnormally thin.

The evocative character of this angiographic pattern should be emphasized because the histological diagnosis is often difficult. The lesions of hepatic fibrosis can escape the needle biopsy. There is often an important discrepancy between the macroscopic and microscopic anatomical patterns; the hepatic biopsy may appear almost normal, while angiography demonstrates a serious atrophic or atropho-hypertrophic transformation of the liver.

Physiopathologically, because these angiographic patterns are similar to those observed during post-necrotic cirrhosis, it is suggested that some of these periportal fibroses can be the sequelae of a previous viral hepatitis. Other authors have reported cases of idiopathic portal hypertension which may be attributed to toxic hepatitis.

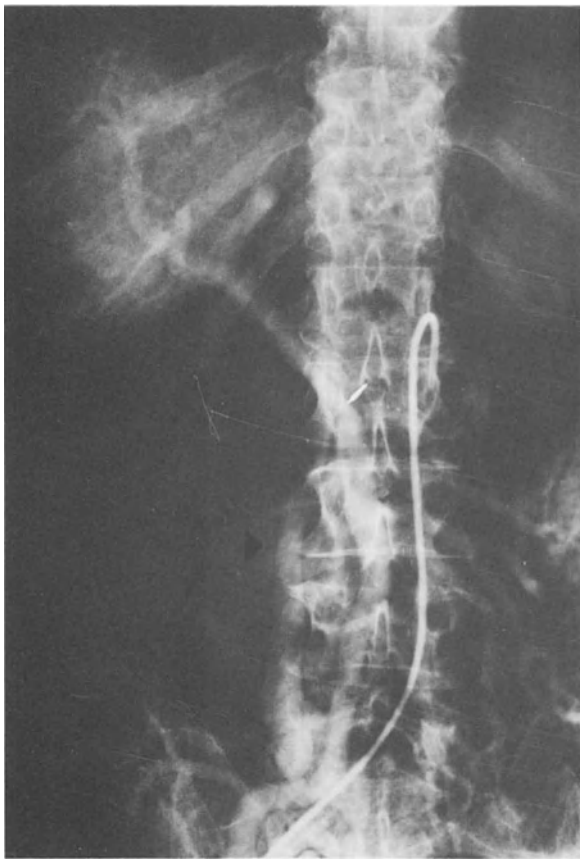
Thus it is reasonable to believe that according to a classical mechanism everywhere else in the organism, this periportal fibrosis is only the late sequela of an inflammatory reaction following various aggressions but principally viral-, toxic-, and drug-related.



33a

Fig. 33. Chronic hepatitis in a 68-year-old female: recurrent jaundice one year after virus hepatitis: Hb As + Liver biopsy: minimal liver cell lesions, steatosis, mild periportal fibrosis (9–9–81).

33a: Normal hepatic artery. The spleen size is within normal limits (14 cm) but the splenic artery is probably some what lengthened and widened. Arterial and venous splenic blood flow seem to be normal.



33b

33b: A relatively good portal flow to the liver is associated with a hepatofugal flow through a wide spontaneous mesenterico-gonadal anstomosis (▲), thus explaining the normal pattern of coeliac arteriography. Recent and minimal intrahepatic obstruction resulting in probably very low portal hypertension owing to the spontaneous porto-systemic shunt.

34a

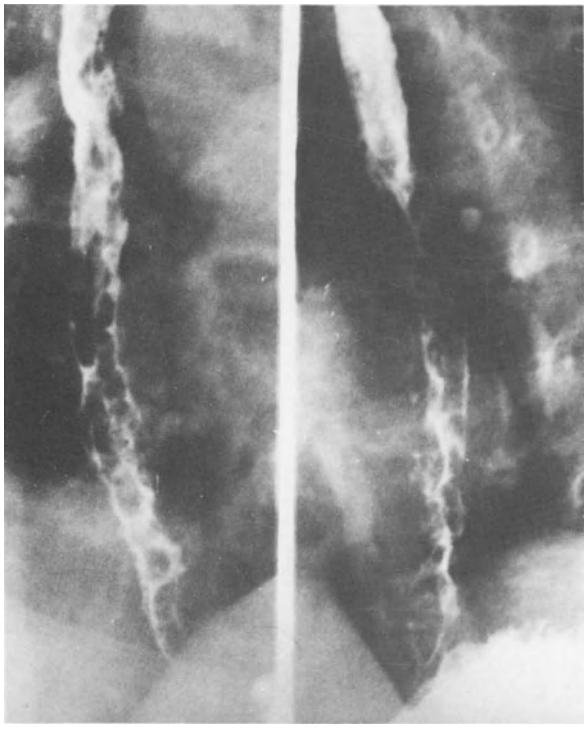
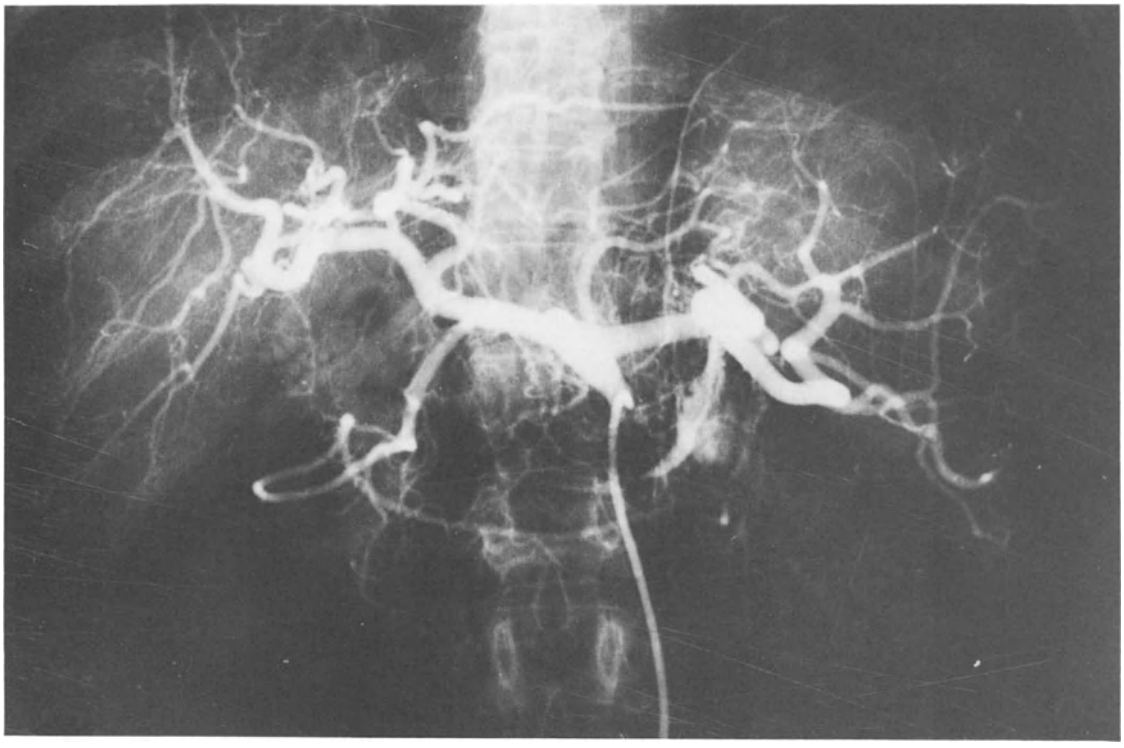


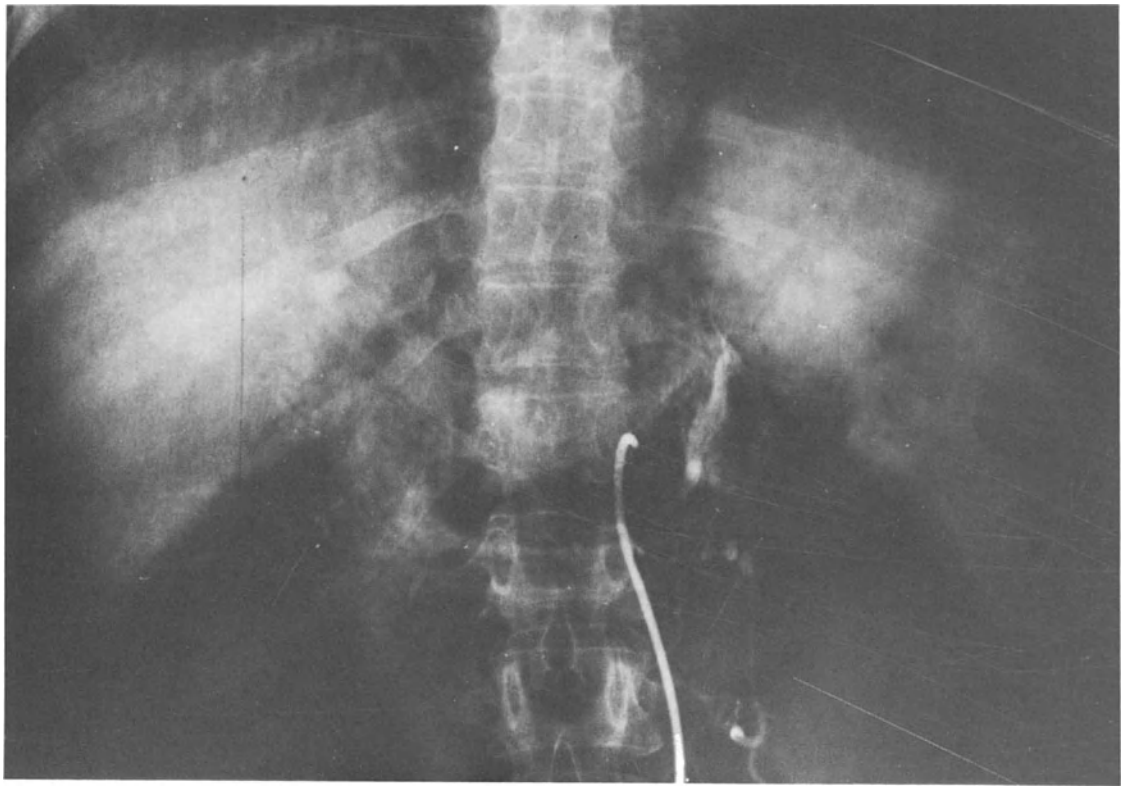
Fig. 34. Alcoholic cirrhosis in a 49-year-old female with massive and recurrent gastro-intestinal bleeding for 2 years (31–5–74).

34a: Large and widespread esophageal varices on Baryum examination.



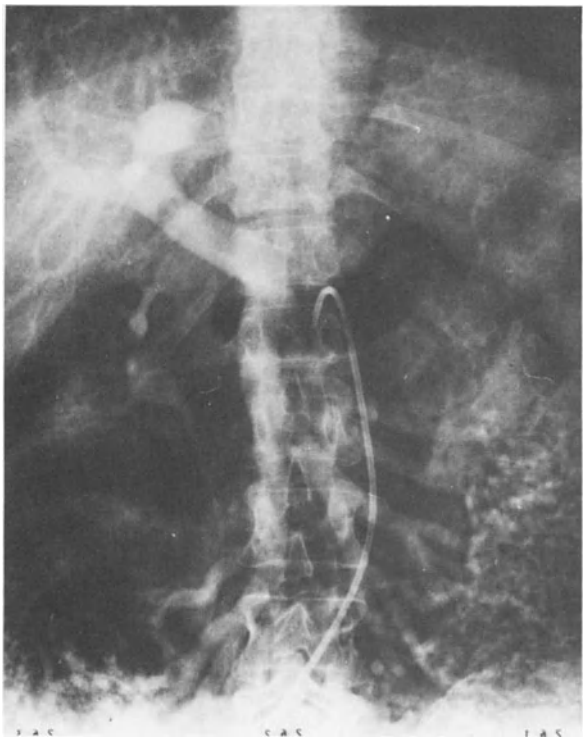
34b

34b: Dilated hepatic artery which signifies the reduced portal inflow.
Moderate splenomegaly (19 cm) with thin intrasplenic arterial branches (inadvertent opacification of the left adrenal gland).



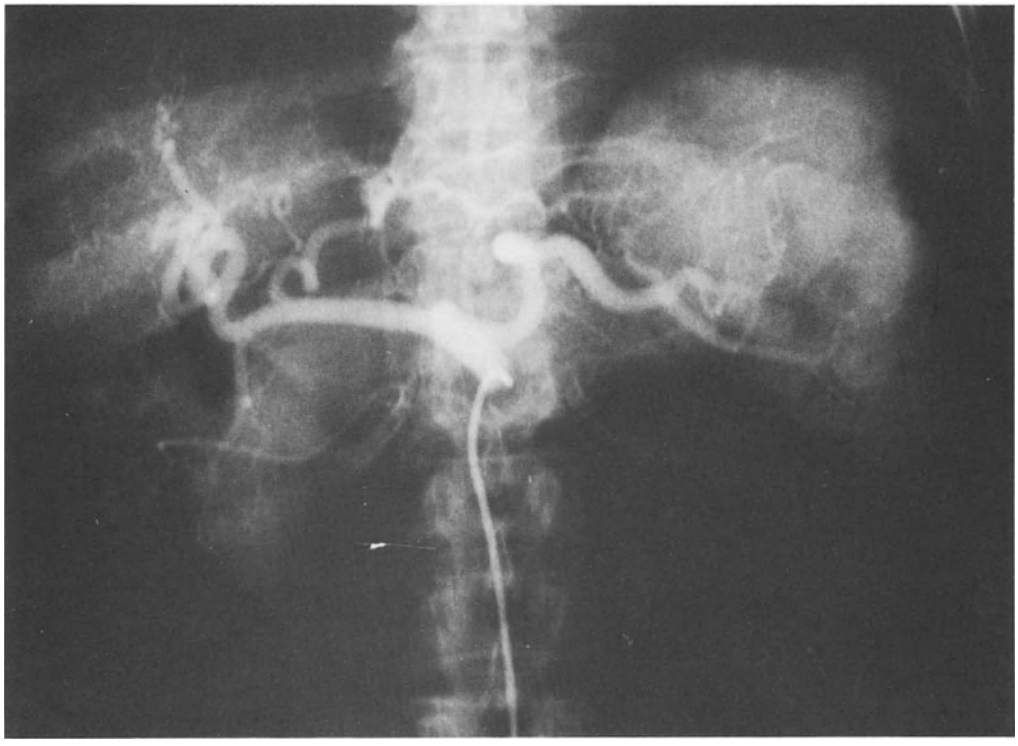
34c

34c: Very faint splenic vein opacification accounting for a pronounced splenic stasis.



34d: Venous phase of S.M.A.: the relatively poor opacification of narrowed intrahepatic portal branches suggests a markedly reduced portal inflow. Neither retrograde opacification of the esophageal varices nor any other hepatofugal way is visualized. As a result, the small intestinal veins are markedly dilated.

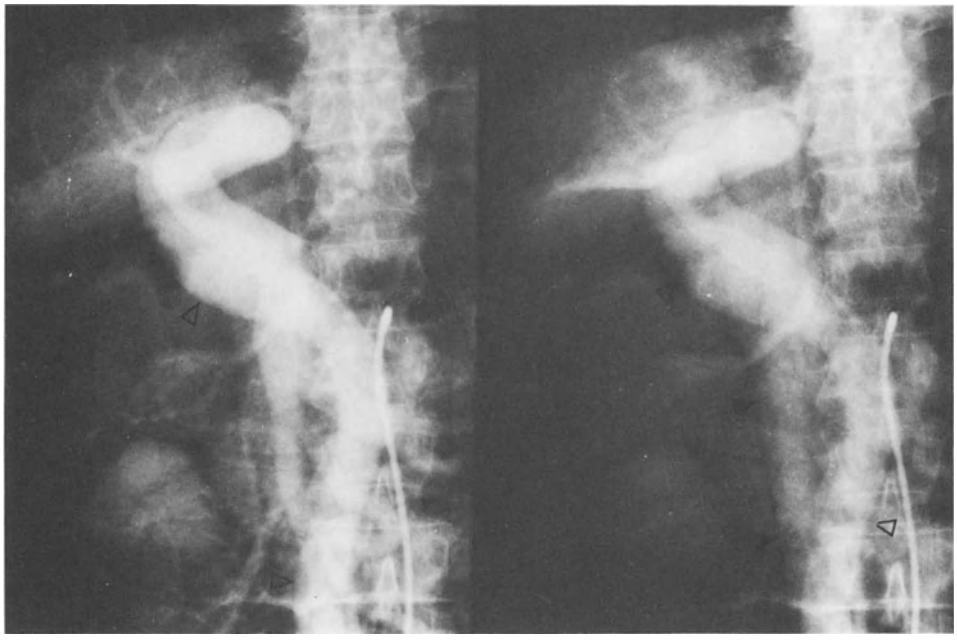
34d



35a

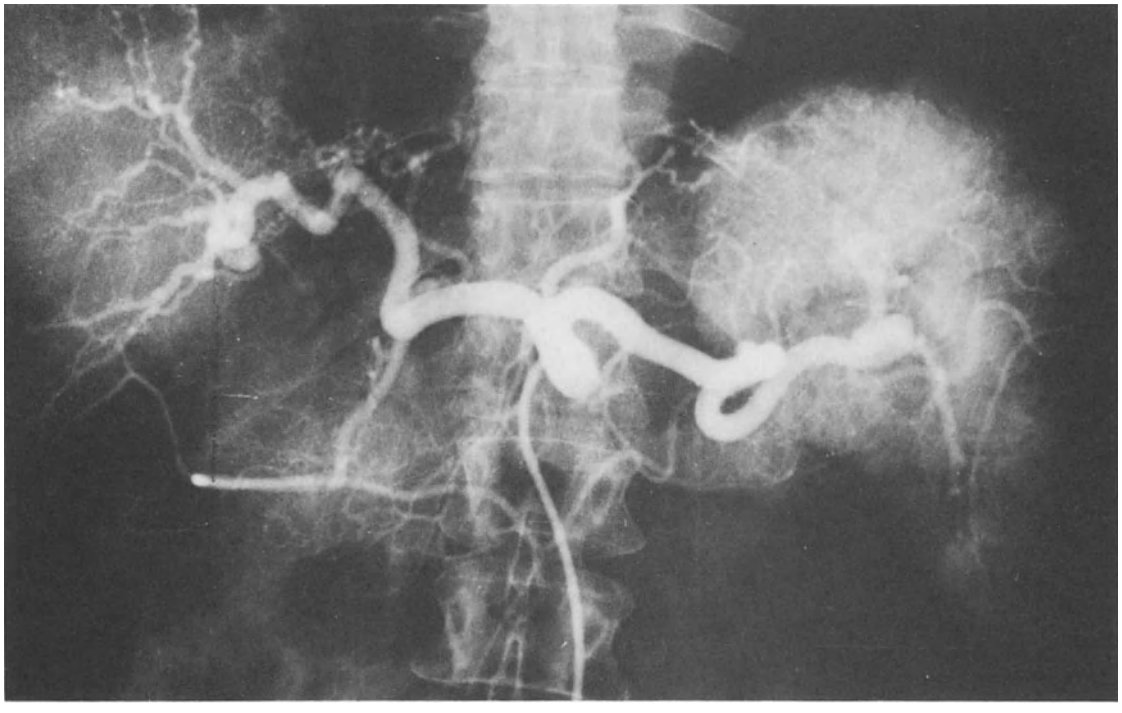
Fig. 35. A 56-year-old female known to have alcoholic cirrhosis with minimal esophageal varices for one year. The patient never bled and was referred for neurologic disorders (28–8–81).

35a: Atrophic liver with markedly dilated hepatic artery which signifies the poor portal inflow. Minimal splenomegaly with early splenography suggesting a normal splenic blood flow rate.



35b

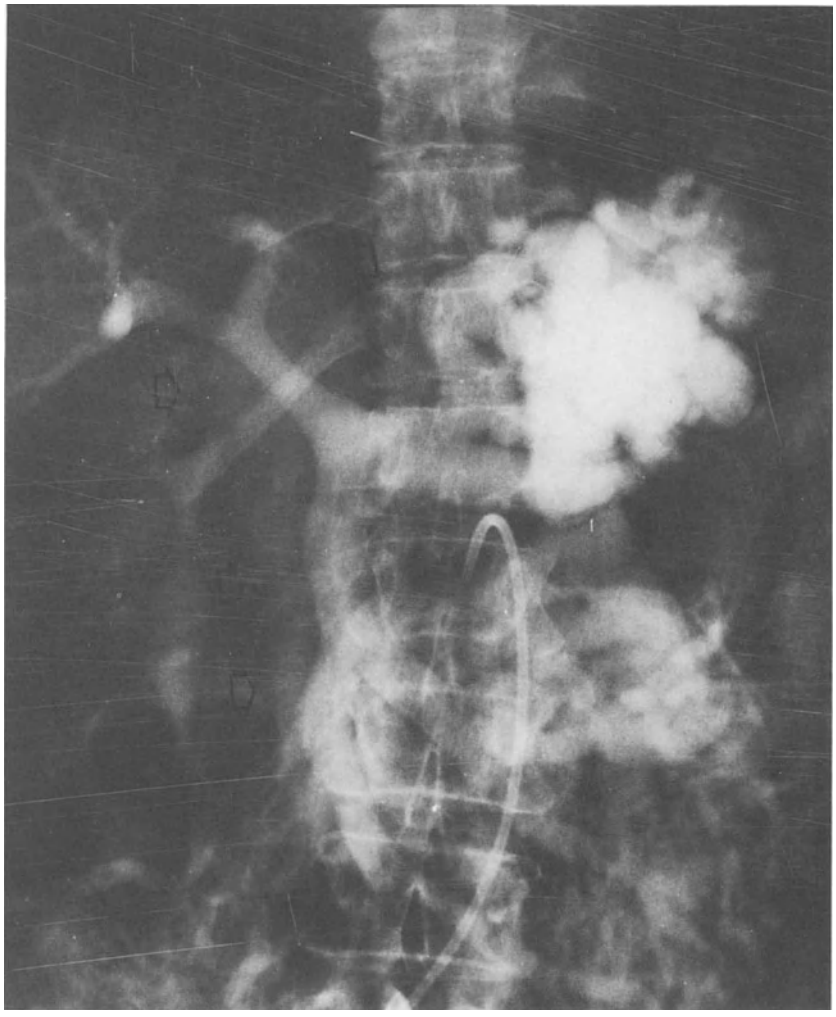
35b: Venous phase of S.M.A.: Very thin right portal vein and intrahepatic portal branches. The portal flow is essentially hepatofugal through a markedly dilated umbilical vein (△) resulting in a dense opacification of the inferior vena cava (▲).



36a

Fig. 36. A 55-year-old female known to have alcoholic cirrhosis with intermittent variceal bleeding ascitis and jaundice for 4 years (21–2–77).
Patient referred for neurologic disorders.

36a: Markedly dilated hepatic artery and minimal splenomegaly (15 cm) with a normal pattern of the splenic blood flow (early dense splenography). Angiographic pattern typical of a markedly decreased portal inflow associated with minimal or moderate portal hypertension.



36b

36b: Venous phase of S.M.A.: Very thinned intrahepatic portal branches. Hepatofugal flow through markedly widened left gastric vein and splenic vein draining towards left renal vein (↑) and inferior vena cava (⌓) via huge gastric varices. Spontaneous spleno-renal anastomosis.

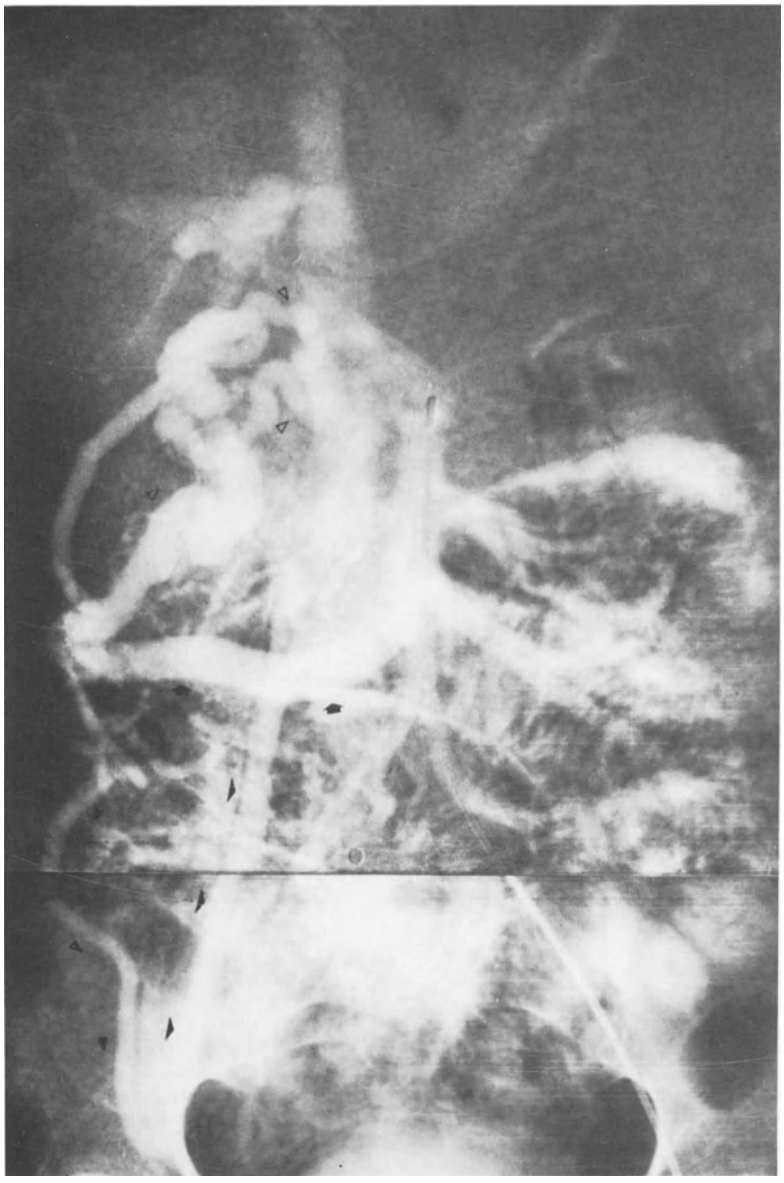
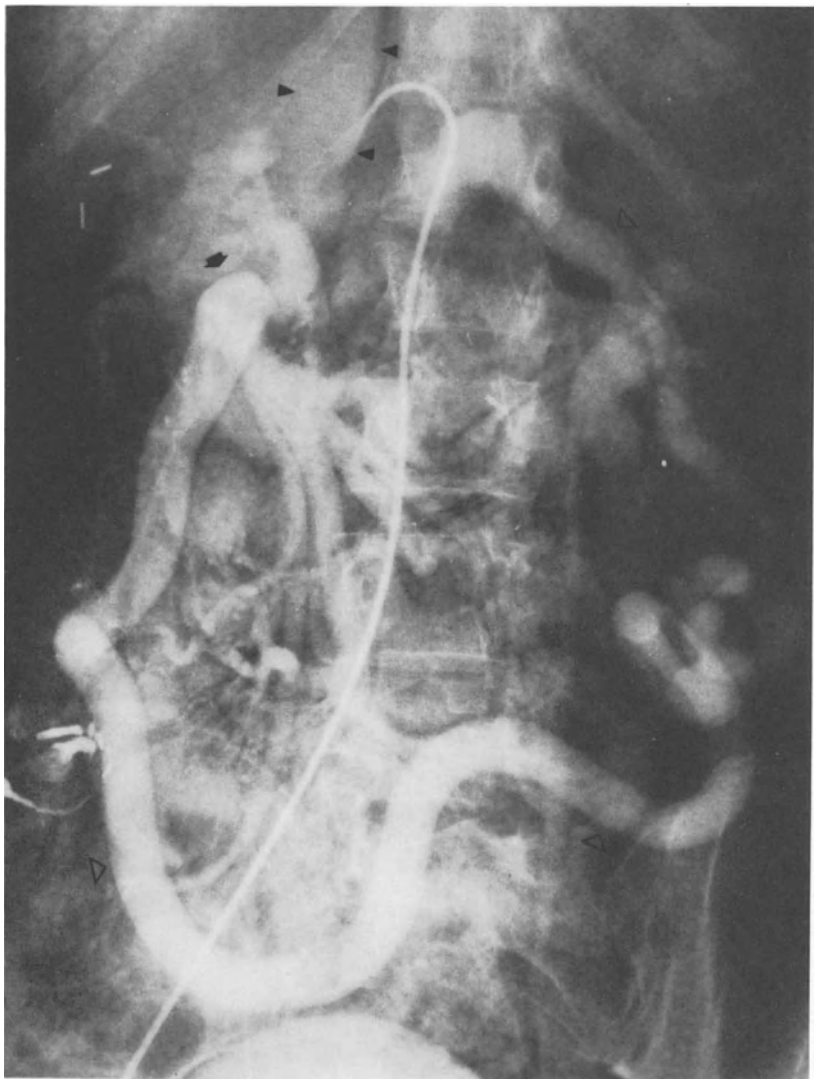


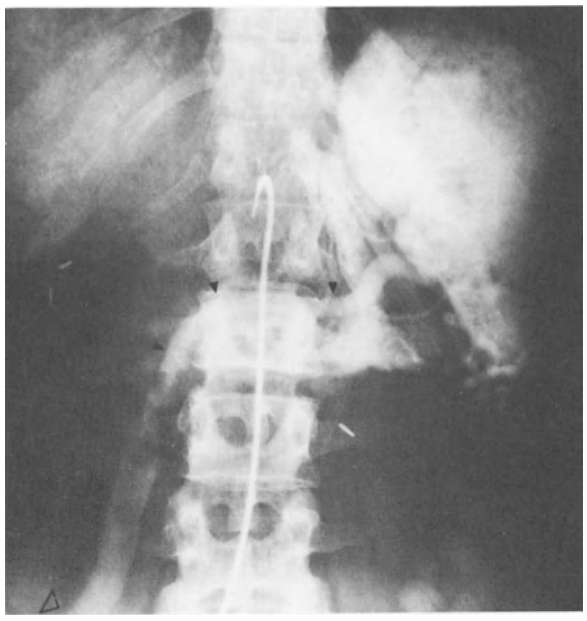
Fig. 37. Alcoholic cirrhosis with spontaneous mesocaval anastomosis in a 42-year-old male.
Venous phase of S.M.A. shows a patent portal vein with a poor hepatopetal flow whereas a dense opacification of the inferior vena cava is evidenced through retroperitoneal anastomosis uniting the dilated right colic vein (↑) to the right gonadal vein (▲) and perirenal veins (△).



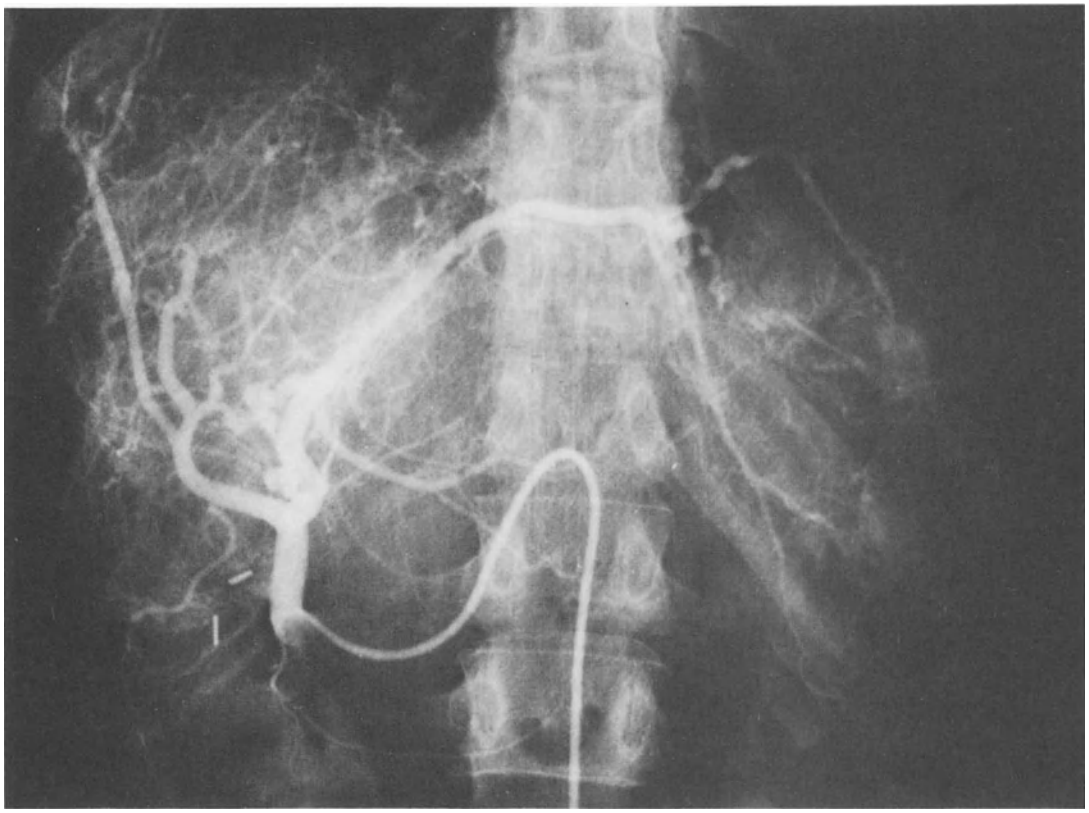
38a

Fig. 38. A 28-year-old healthy female thought to have one hepatic tumor in the right lobe and another one in the left lobe on the basis of ultrasound examination performed for mild epigastric pain. No sign of portal hypertension.
Surgical biopsy suggests either focal nodular hyperplasia or liver adenoma-Normal function liver tests except alkaline phosphatasis (1000 u.i).
38a: Venous phase of S.M.A. (17–5–84).
Superior mesenteric vein seems to be completely interrupted (↑). Totally hepatofugal flow through a wide retroperitoneal vein probably originating from middle colic vein (△) and resulting in opacification of left renal vein and inferior vena cava (▲).
This angiographic pattern can not be explained by portal vein thrombosis as there is no hepatopetal collateral flow. In fact, portal vein was proved to be patent during surgical exploration. A functionnal obstruction of the portal vein associated with an intrahepatic obstacle is then likely.

38b: Venous phase of coeliac arteriography: the splenic venous outflow (▲) also drains through the meso-renal anastomosis (△) together with the superior mesenteric venous blood.

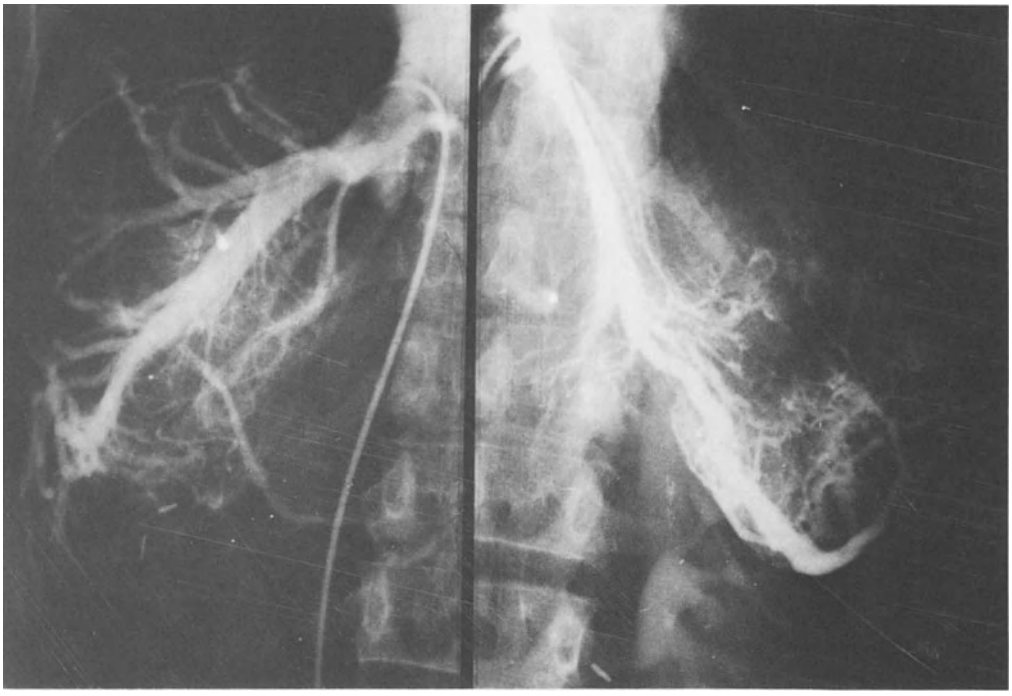


38b



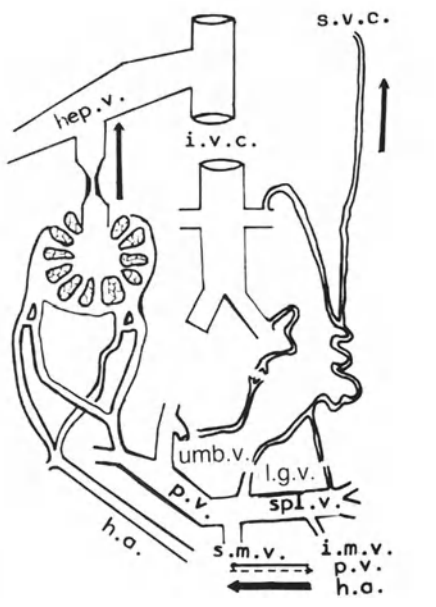
38c

38c: Arterial widening compensating for the lost portal flow. Hypertrophic left lobe and segment 8 without tumoral pattern associated with a marked atrophy of the segments 5, 6 and 7 of the right lobe.

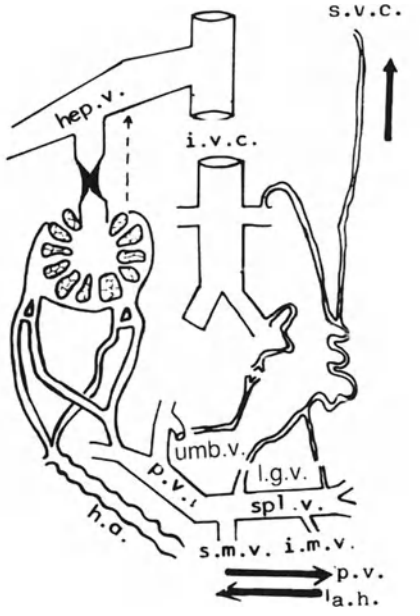


38d

38d: Hepatic venography (5–6–84).
The lack of tumoral pattern is confirmed.
Wedged hepatic pressure: 10 mm Hg. Pseudo-tumoral regenerative modules associated with a presinusoïdal obstruction seems more likely than true hepatic tumors.



39a

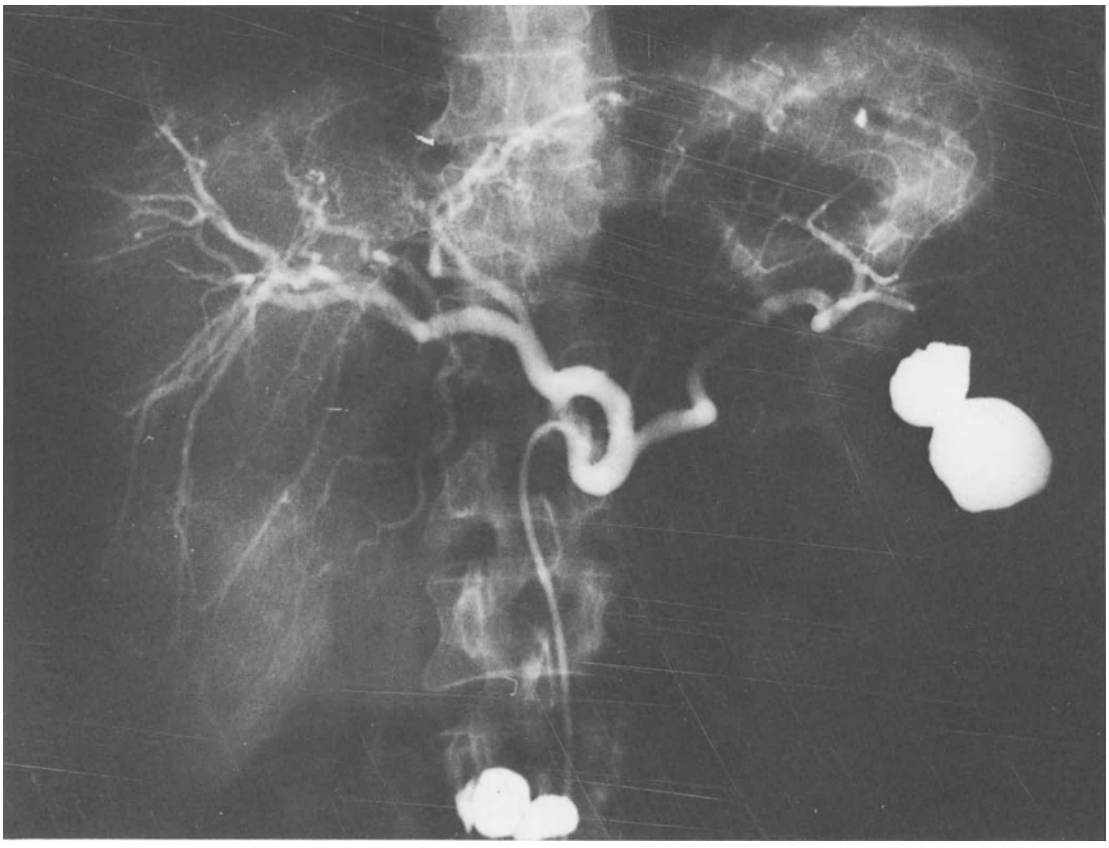


39b

Fig. 39: Schematic drawing of the hepatic circulation in case of reversal of intrahepatic portal flow.

39a: Partial reversal of the intrahepatic portal flow. Because of a relatively low hepatofugal flow rate a bidirectional flow in the portal vein itself can be documented by arterio-portography.

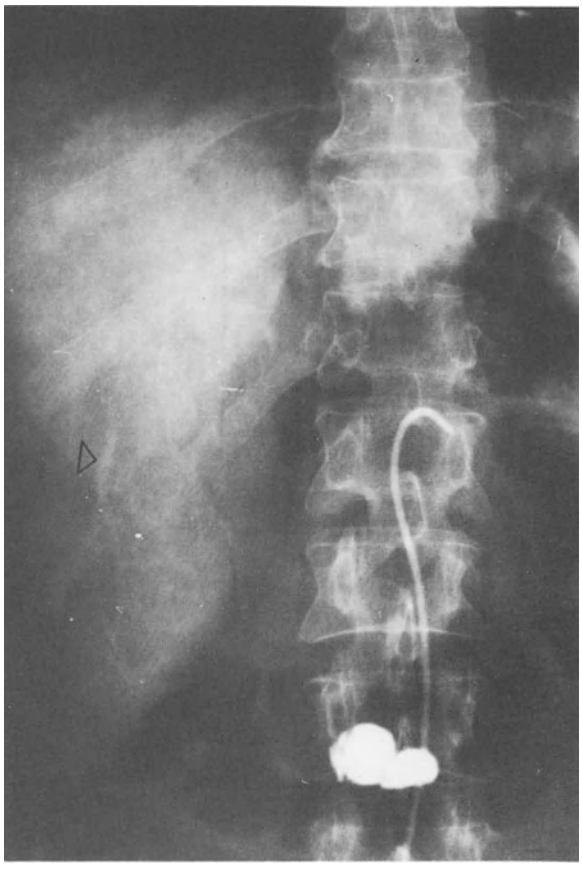
39b: Complete reversal of the intrahepatic portal flow.
A more severe post-sinusoïdal obstruction results in a completely hepatofugal flow through the extrahepatic portal vein.



40a

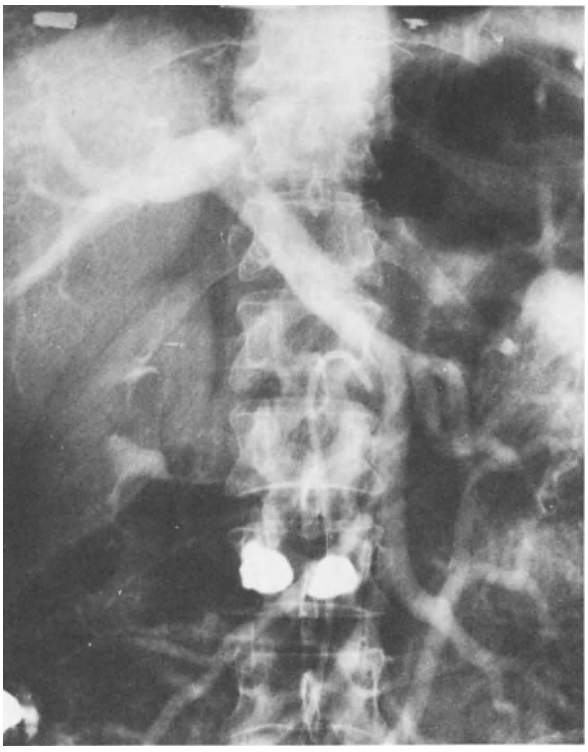
Fig. 40: Partial reversal of the intrahepatic portal flow in a 47-year-old alcoholic patient referred for epigastric pain thought to be due to pancreatitis (3–1–77).

40a: Coeliac arteriography: Normal sized spleen (14 cm) with thinned intrasplenic arterial branches which signifies a slowed intrasplenic blood flow.
Hepatomegaly and dilated hepatic artery which signifies the reduced portal inflow.



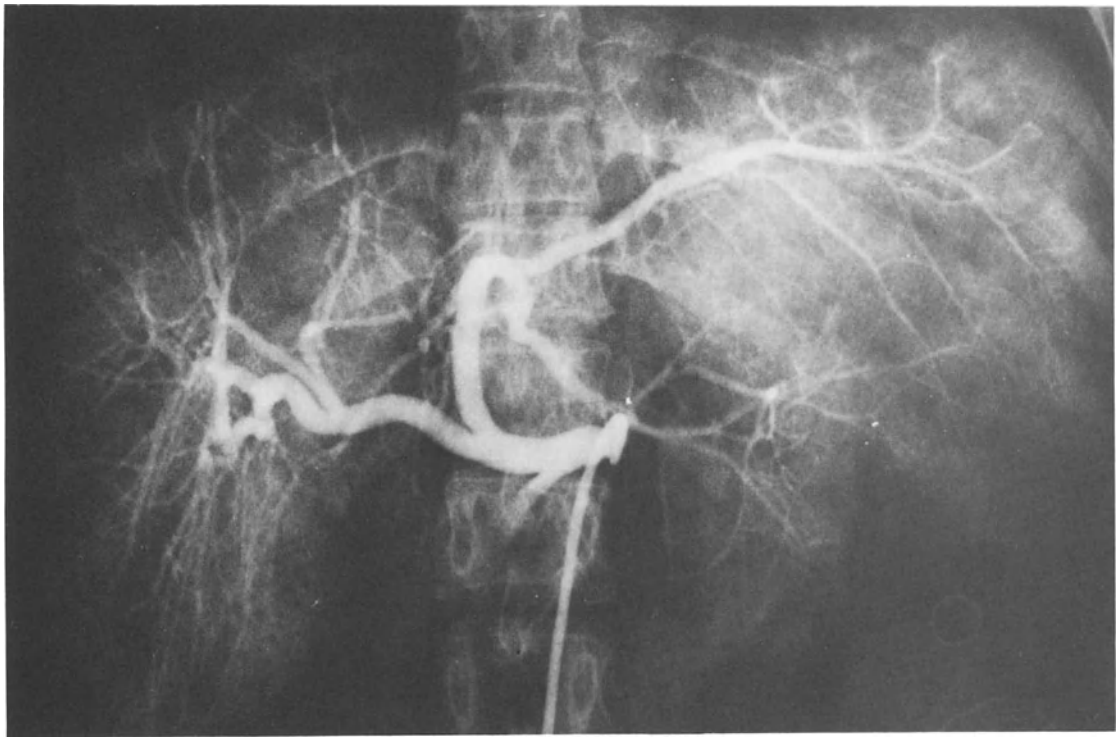
40b

40b: 7 seconds later the splenic vein is not opacified whereas a dense arterial hepatography is visualized as well as retrograde filling of a small intrahepatic portal branch (△).



40c

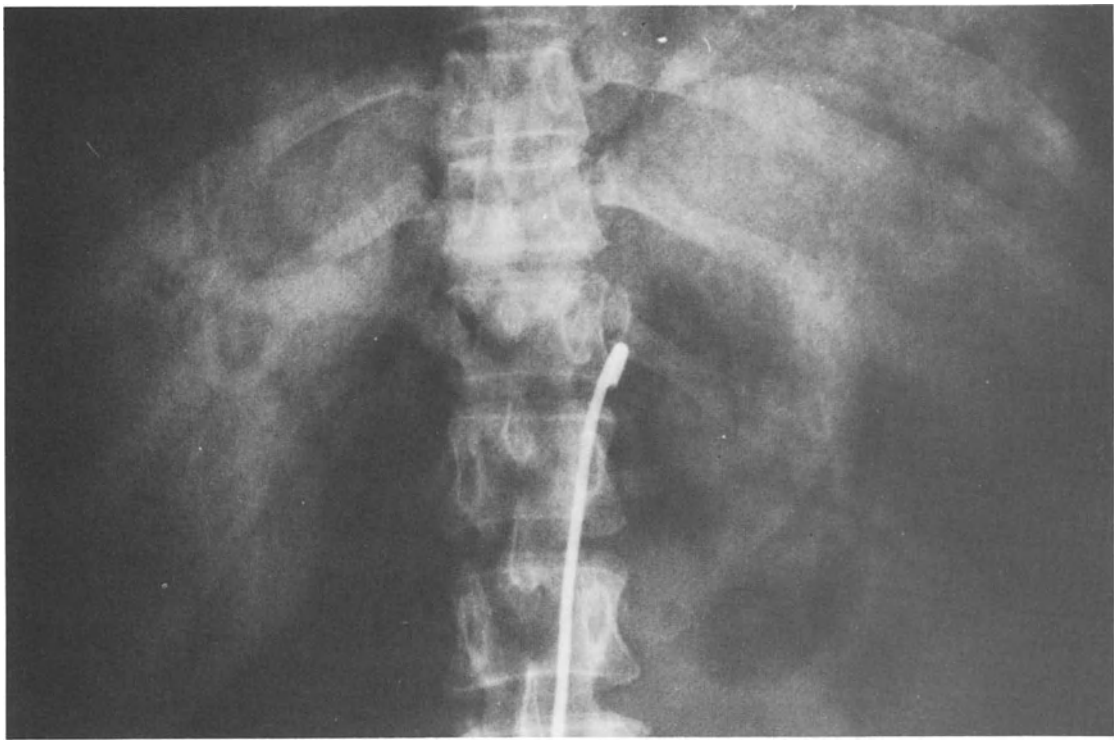
40c: Venous phase of S.M.A.: Hepatofulgal flow in the inferior mesenteric vein. Narrowed and poorly opacified intrahepatic portal branches with no portal opacification in the lower part of the right lobe where the intrahepatic portal flow was shown to be reversed (Segment 5).
Angiographic pattern of a severe and quickly progressive intrahepatic obstruction. Liver biopsy (12–1–77): Highly evolving cirrhosis.



41a

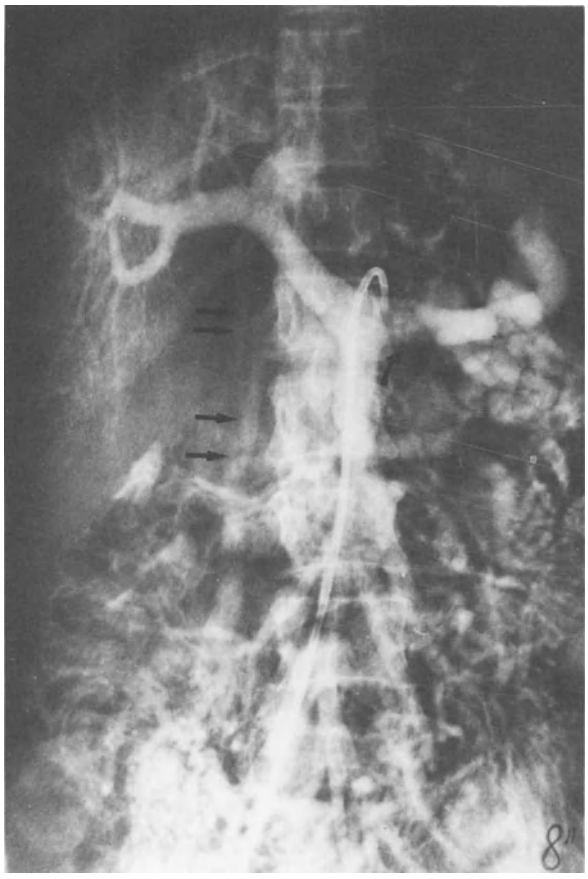
Fig. 41. Cirrhosis probably of the post-hepatitis type in a 40-year-old female with reversed intrahepatic portal flow (no history of alcohol intake – virus hepatitis 20 years earlier) (16–9–75).

41a: Selective hepatic arteriography: hepatomegaly with marked dilatation of hepatic artery.

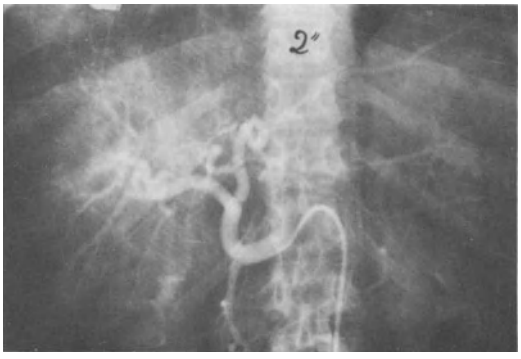


41b

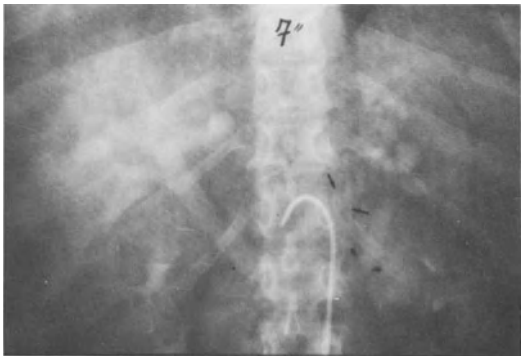
41b: Retrograde opacification of the portal vein clearly demonstrated at the 8th second.



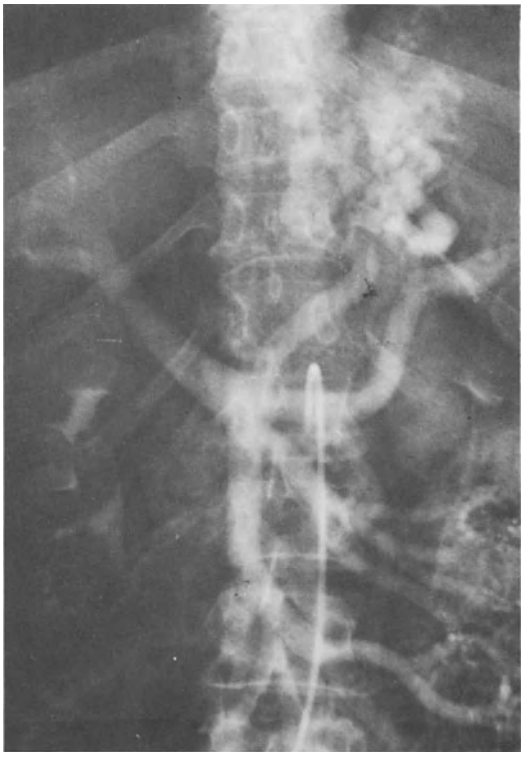
41c



42a



42b



42c

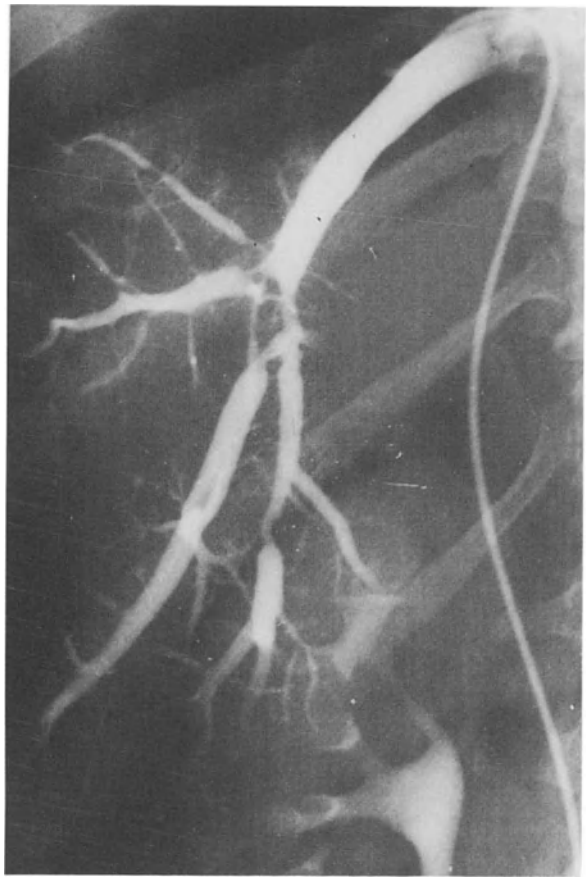
41c: Venous phase of S.M.A.: opacification of the portal vein together with few thin intrahepatic branches (bidirectional flow).
Predominant hepatofugal flow through splenic vein and right gonaldal vein (↑) (Spontaneous meso-gonadal anastomosis).

Fig. 42. Alcoholic cirrhosis known for one year in a 36-year-old female. Diagnosis confirmed by laparoscopy and liver biopsy (very dense collagen sclerosis) (14–9–76).

42a: Selective hepatic arteriography (2nd sec): markedly dilated hepatic artery with early dense opacification of some areas of the right liver.

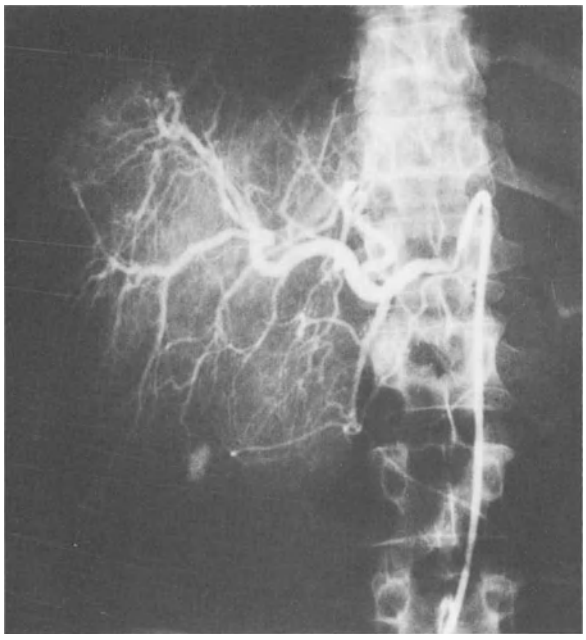
42b: Retrograde opacification of the portal vein at the 7th second with left gastric vein and splenic vein faintly visualised, which signifies the completely hepatofugal flow through portal vein.

42c: Venous phase of S.M.A.: progressive weakening of portal vein opacification with no hepatopetal flow (functional obstruction).
Essentially hepatofugal flow through widened splenic vein, left gastric vein and varices.



42d

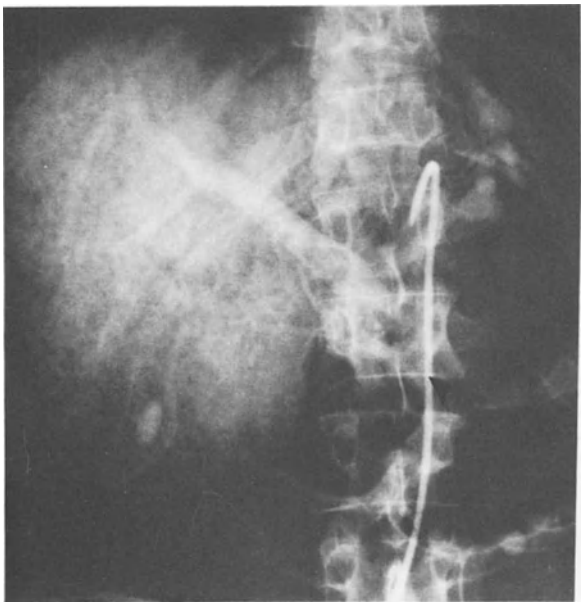
42d: Hepatic venography: multiple stenosis of the right hepatic vein branches. Wedged hepatic venography: 22 cm saline.



43a

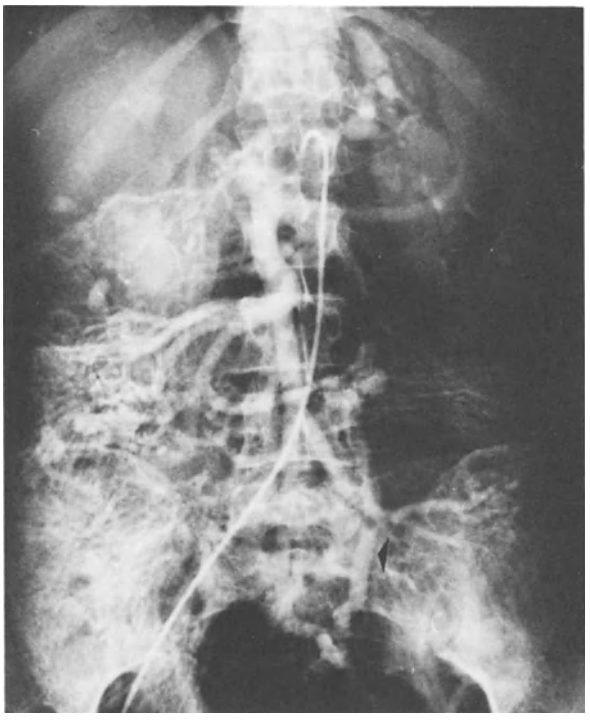
Fig. 43. Alcoholic cirrhosis with complete reversal of the intrahepatic portal flow in a 48-year-old female, referred for lower intestinal bleeding (10–7–80). (Total colectomy for ulcerative colitis 10 years previously.)

43a: Selective hepatic arteriography: markedly dilated hepatic artery resulting in dense arterial hepatography.

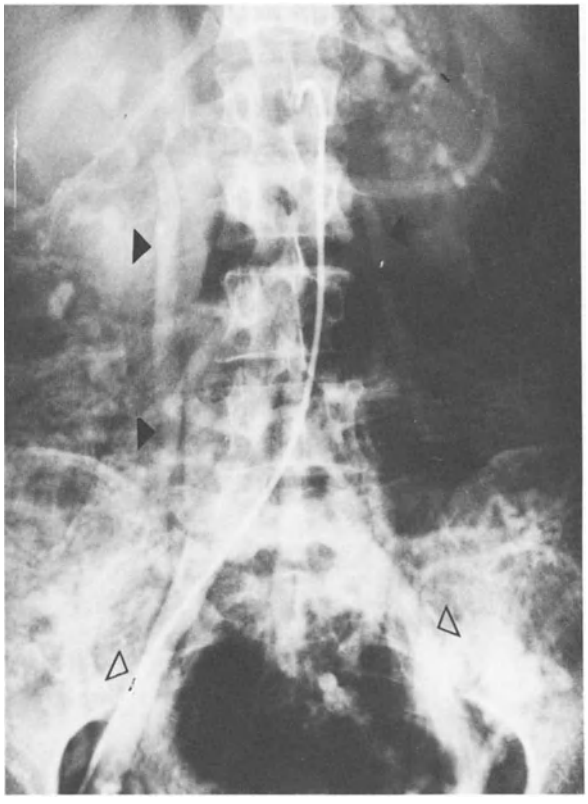


43b

43b: Retrograde opacification of the portal vein, then the left gastric vein (↑) and the inferior mesenteric vein (↑) 5 sec later, which signifies the totally hepatofugal portal flow.

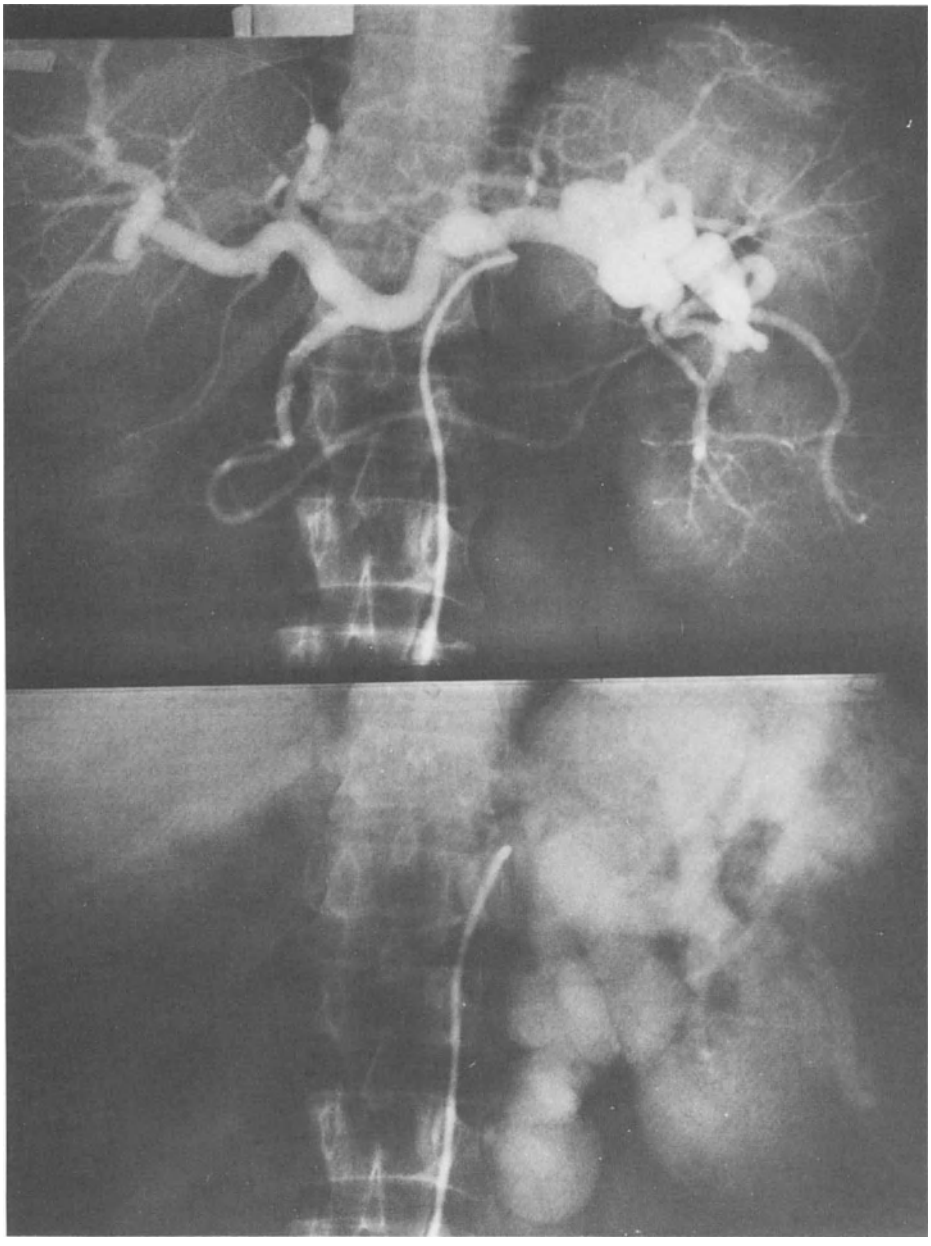


43c



43d

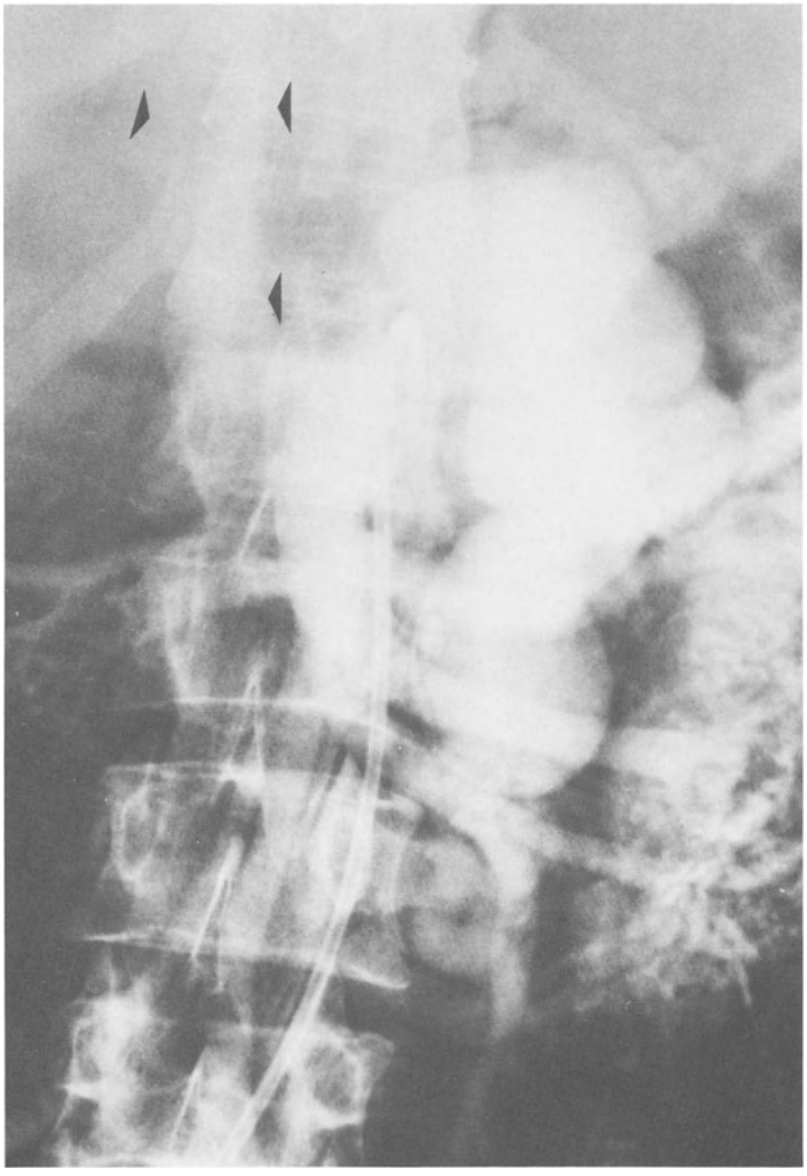
43c–43d: Venous phase of S.M.A.: functional obstruction of the portal stem and retrograde flow in the inferior mesenteric vein and intestinal varices (↑) draining toward right and left iliac veins (△) and inferior vena cava through both gonadal veins (▲).



44a

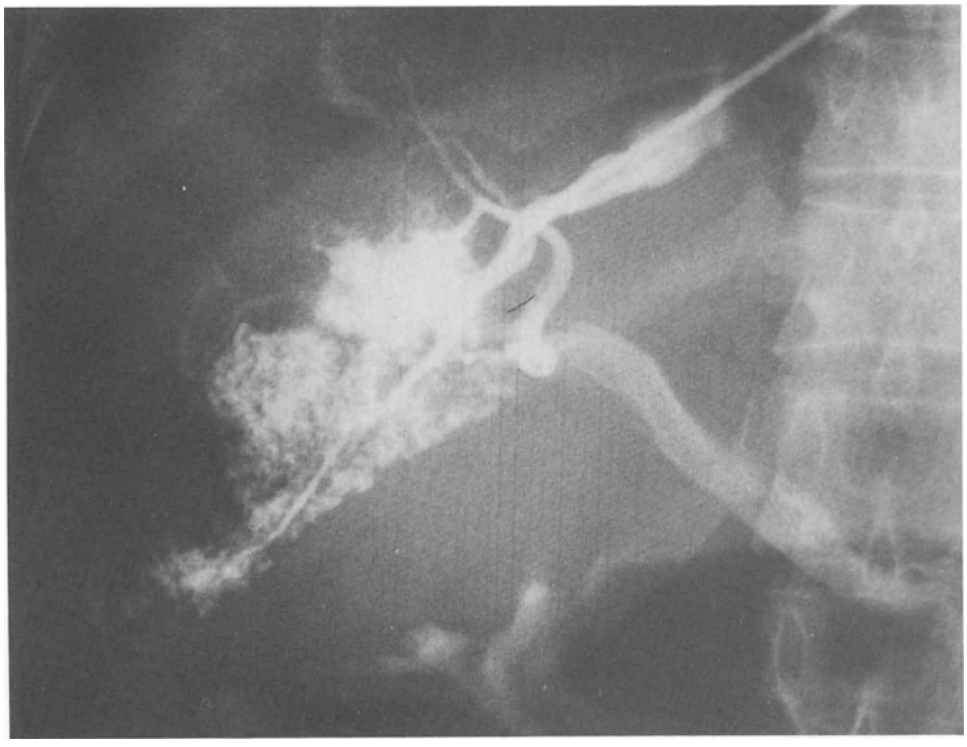
Fig. 44. Cirrhosis of unknown origin in a 38-year-old male referred for intermittent jaundice, weight loss, diabetes, hemochromatosis and mild liver function test abnormalities (26–3–76).

44a: Markedly widened hepatic artery suggesting a very low hepatopetal flow. Splenomegaly (19 cm) with widened and lengthened splenic artery.
Splenic vein draining through a spontaneous spleno-renal anastomosis.



44b

44b: Venous phase of S.M.A.: Portal vein not visualized. The superior mesenteric vein drains through the wide and tortuous spleno-renal anastomosis towards the inferior vena cava (▲).



44c

44c: Wedged hepatic venography: marked retrograde filling of the portal vein, which signifies the hepatofugal portal flow responsible for the lack of portal vein opacification on the venous phase of S.M.A.
In spite of the totally hepatofugal portal flow this patient is still doing well 6 years later (26–5–82) and experienced neither digestive bleeding nor ascistis.

45a

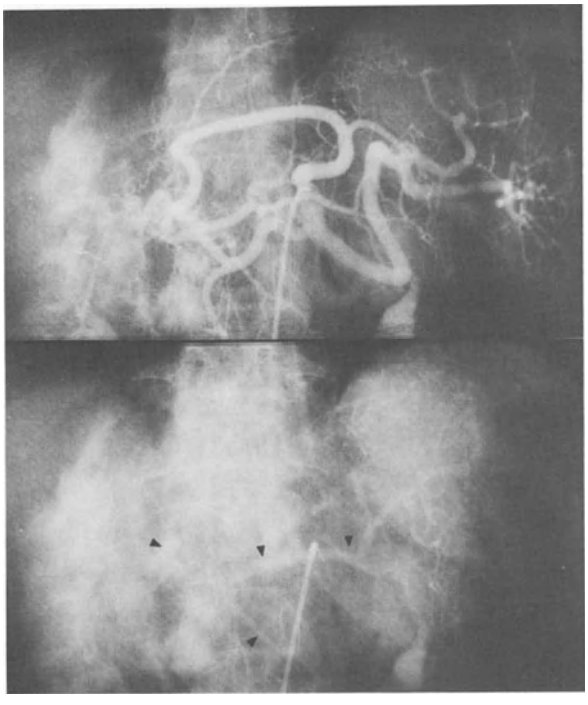
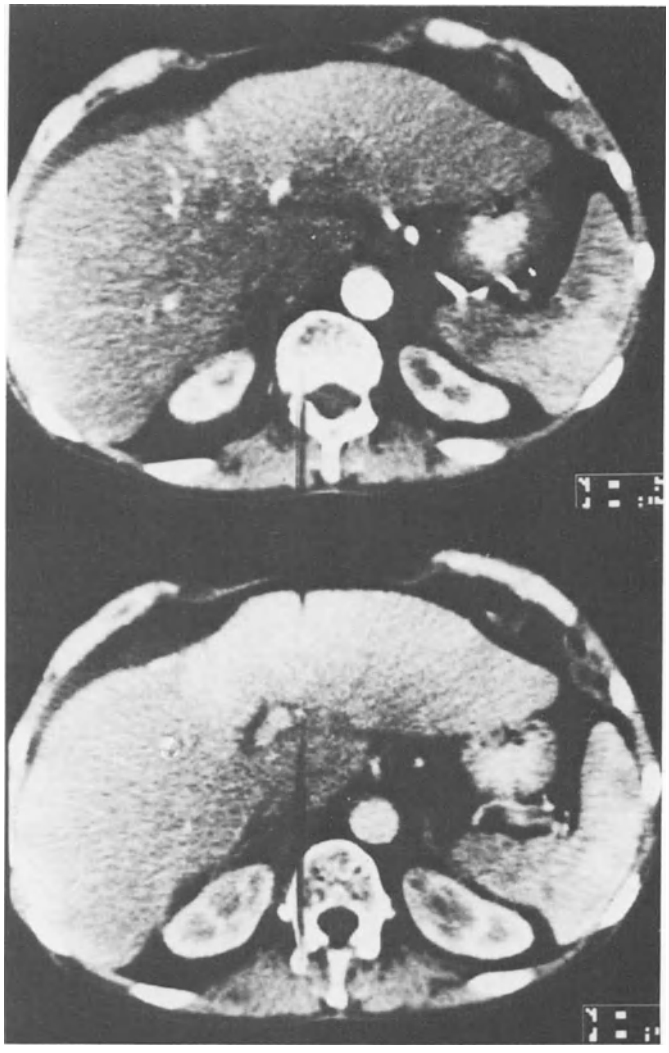
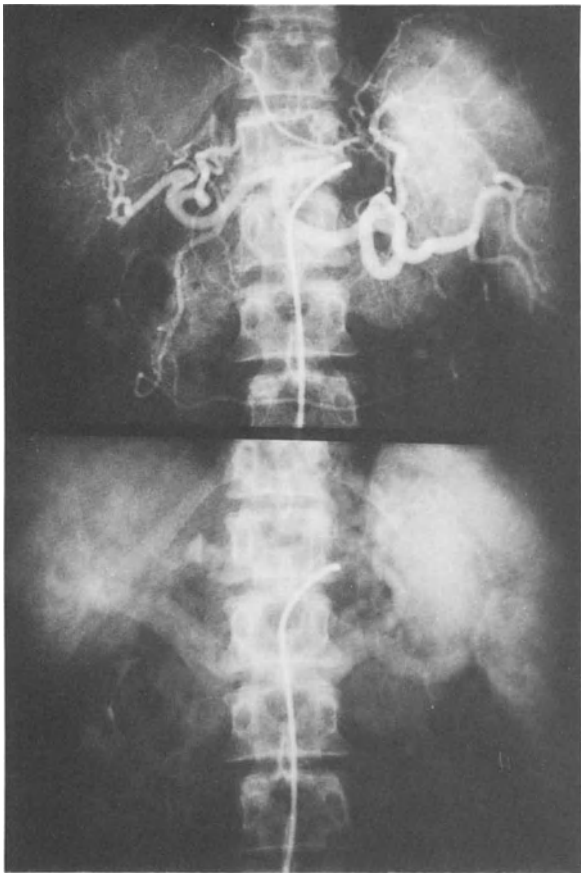


Fig. 45. Alcoholic cirrhosis with partial reversal of the intrahepatic portal flow in a 52-year-old male.
45a: Coeliac arteriography (1–8–82).
Splenomegaly (18 cm) with thin intrasplenic arterial branches and poor opacification of the splenic vein on the late films.
Dilated left hepatic artery with dense arterial hepatography in the segment 4 and early retrograde filling of the left portal branches (5th sec) (▲).

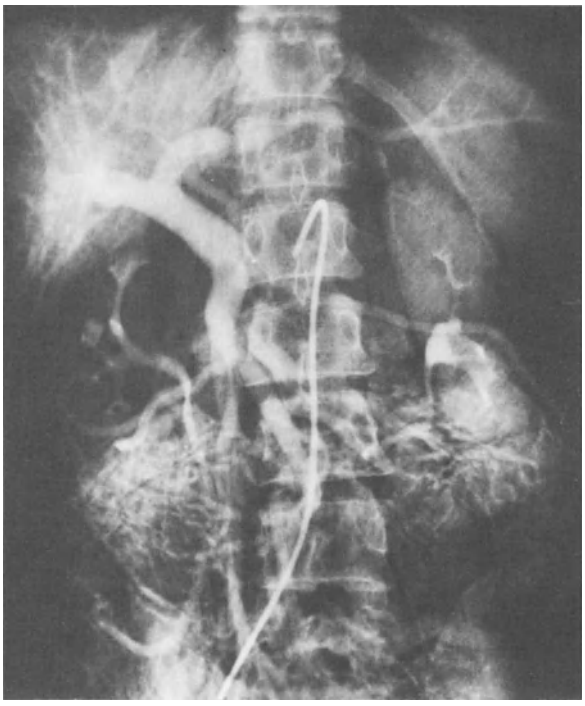


45b

45b: Angio C.T. (12–8–82). On arterial phase the dilated left hepatic artery is visible (↑) as well as several intrahepatic vessels in the segment 4 corresponding to the arterio-portal shunting.
20 seconds later, portal vein is seen in the hepatic hilum and portal hepatography as well.



46a

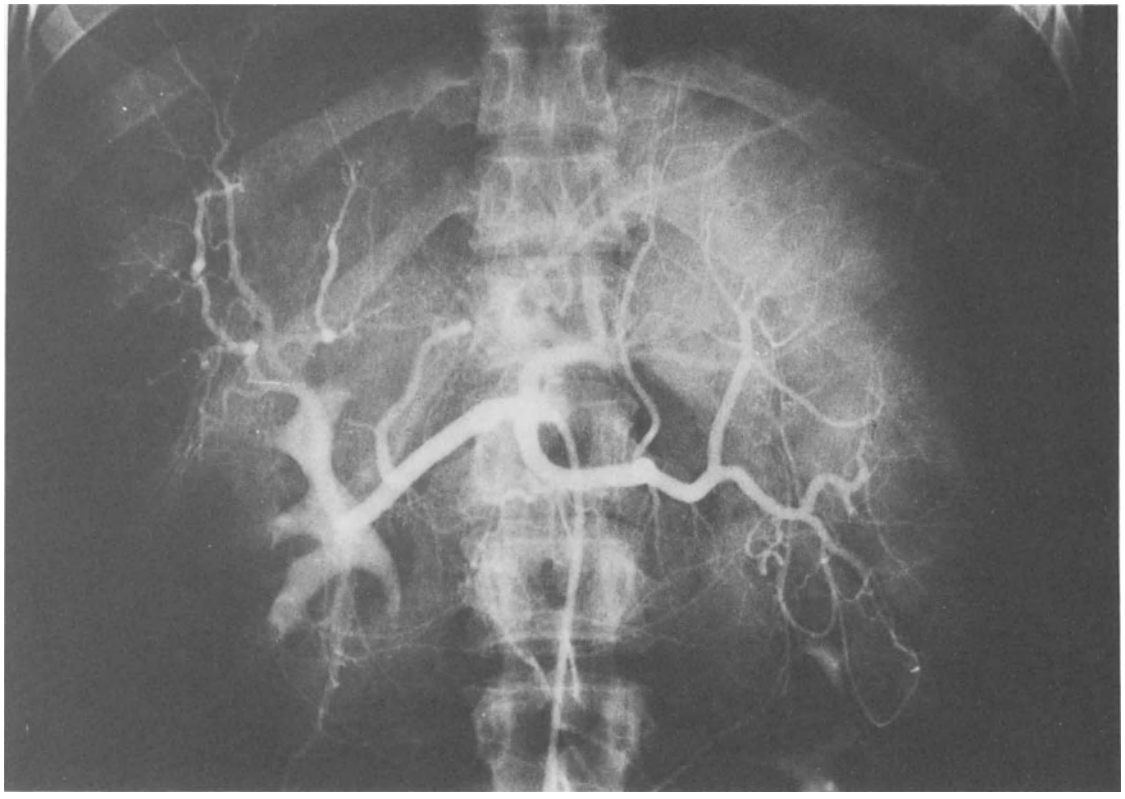


46b

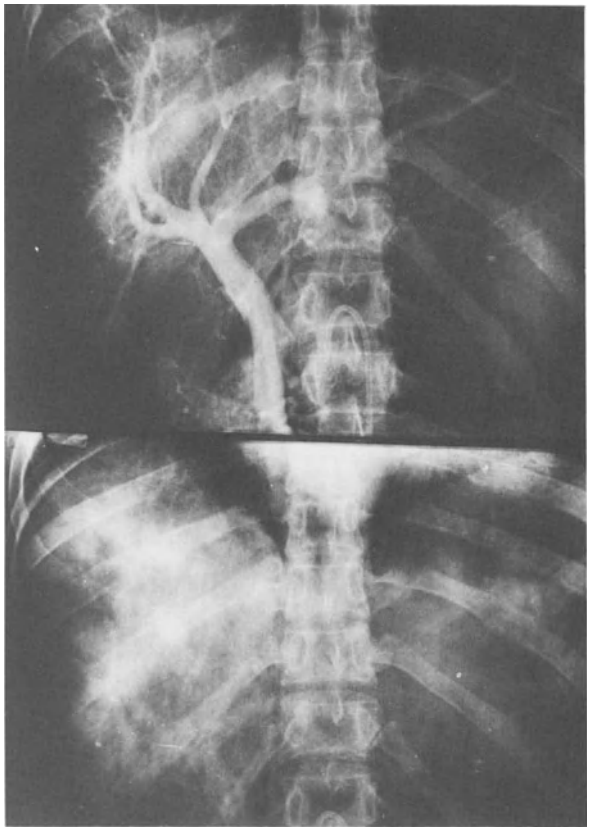
Fig. 46. Slowly progressive post-hepatitis cirrhosis in a 53-year-old female.
Several episodes of jaundice for 30 years.
First variceal bleeding 2 years ago.
Hypersplenism (13–5–81).

46a: Pronounced splenomegaly (20 cm) with normal splenography, visualization of the splenoportal axis and faint opacification of gastric varices through a short gastric vein. Slightly dilated hepatic artery with normal intrahepatic branches.

46b: Venous phase of S.M.A.: Normal mesenterico-portal axis with realively good opacification of slightly narrowed intrahepatic portal branches evidencing an atrophic right lobe associated with a hypertrophic left one.
Minimal retrograde filling of the inferior mesenteric vein. Angiographic pattern suggesting a moderate and slowly progressive intrahepatic obstruction.



47a



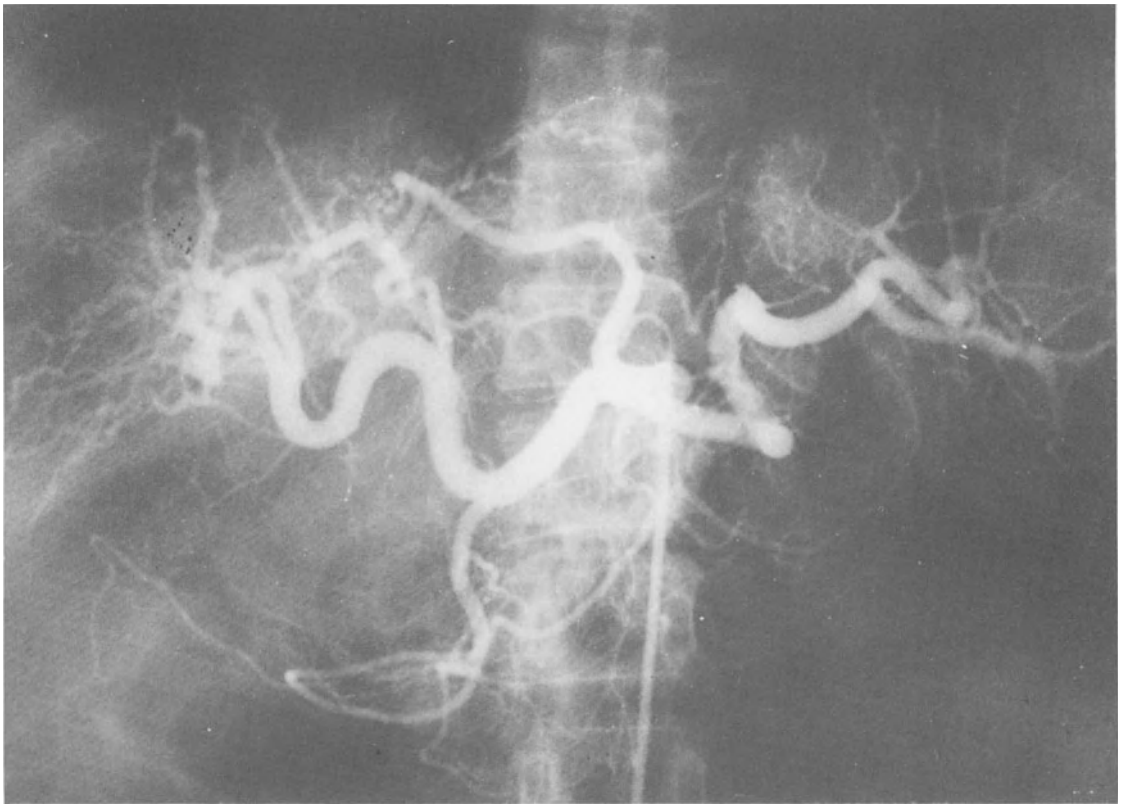
47b

Fig. 47. Alcoholic cirrhosis with heterogeneous intrahepatic portal flow in a 33-year-old female referred for ascitis and esophageal varices bleeding (25–10–77).

47a: Splenomegaly (19 cm) and moderate widening of some intrahepatic arterial branches.

47b: Venous phase of S.M.A.: good visualization of several intrahepatic portal branches resulting in dense opacification of some areas of the liver parenchyma whereas other territories are not opacified, so that a heterogeneous portal hepatography is seen on the late films. Hepatofugal collateral ways are not visualized.

Following a meso-caval interposition shunt (12/77) the patient remains well 6 years later.



48a

48b

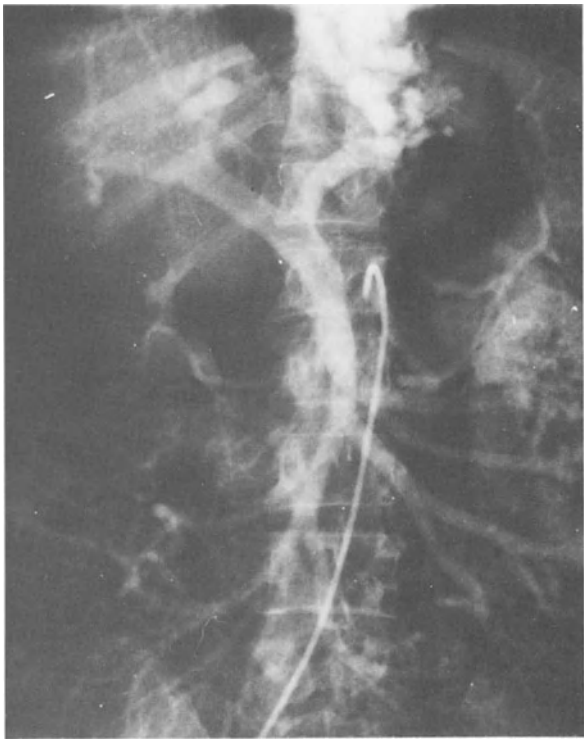
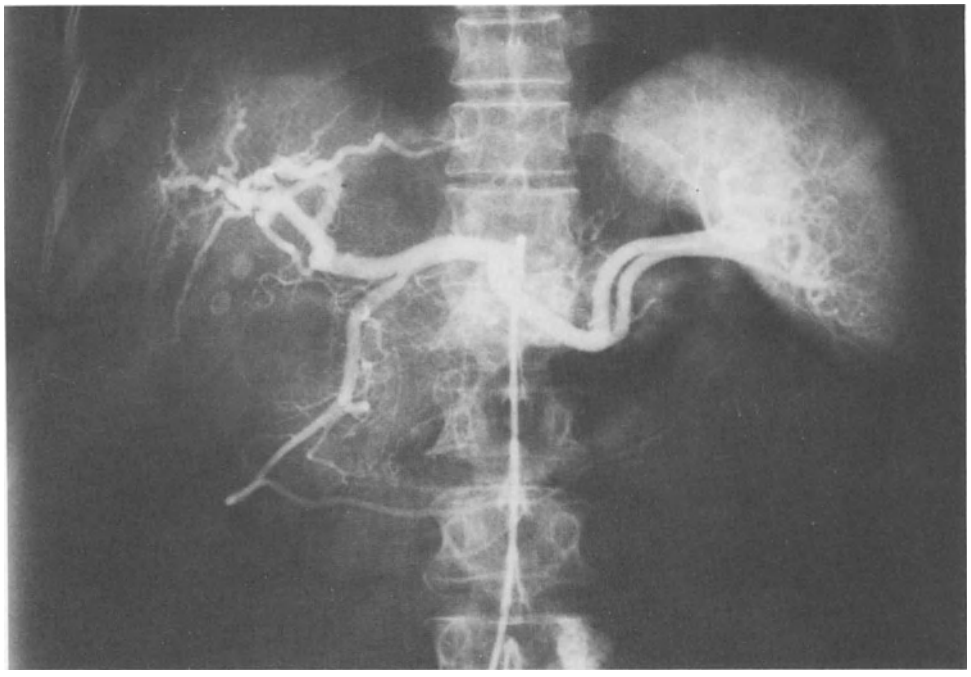


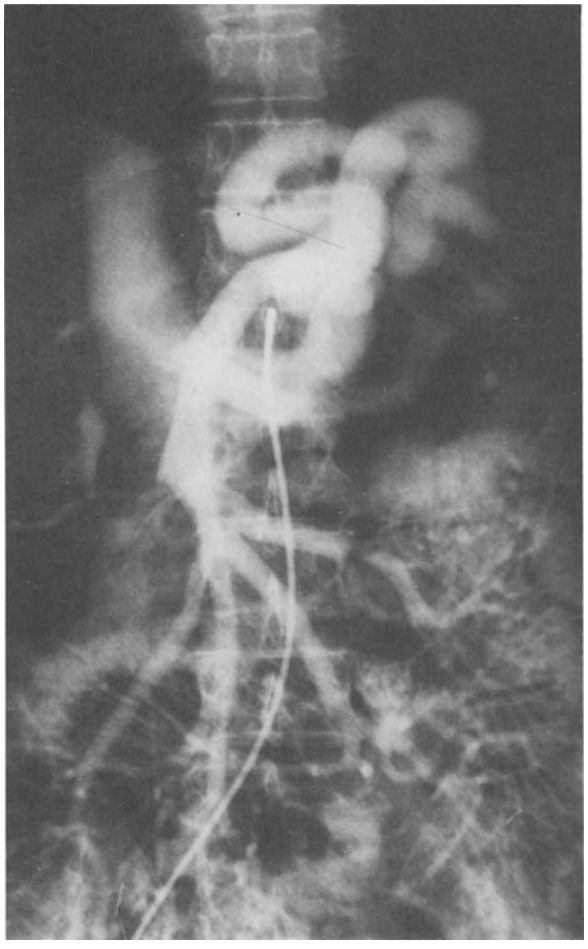
Fig. 48. Alcoholic cirrhosis with severe obstruction to the portal inflow in a 51-year-old female (20–12–82).
Portal pressure: 30 mm Hg.

48a: Minimal splenomegaly (15 cm) with thin intrasplenic arterial branches (low flow rate pattern).
Atrophic liver with markedly dilated hepatic artery which signifies the greatly reduced portal inflow.

48b: Venous phase of S.M.A.: very poor filling of portal vein and few intrahepatic branches.
Dense opacification of a wide left gastric vein and esophageal varices.



49a

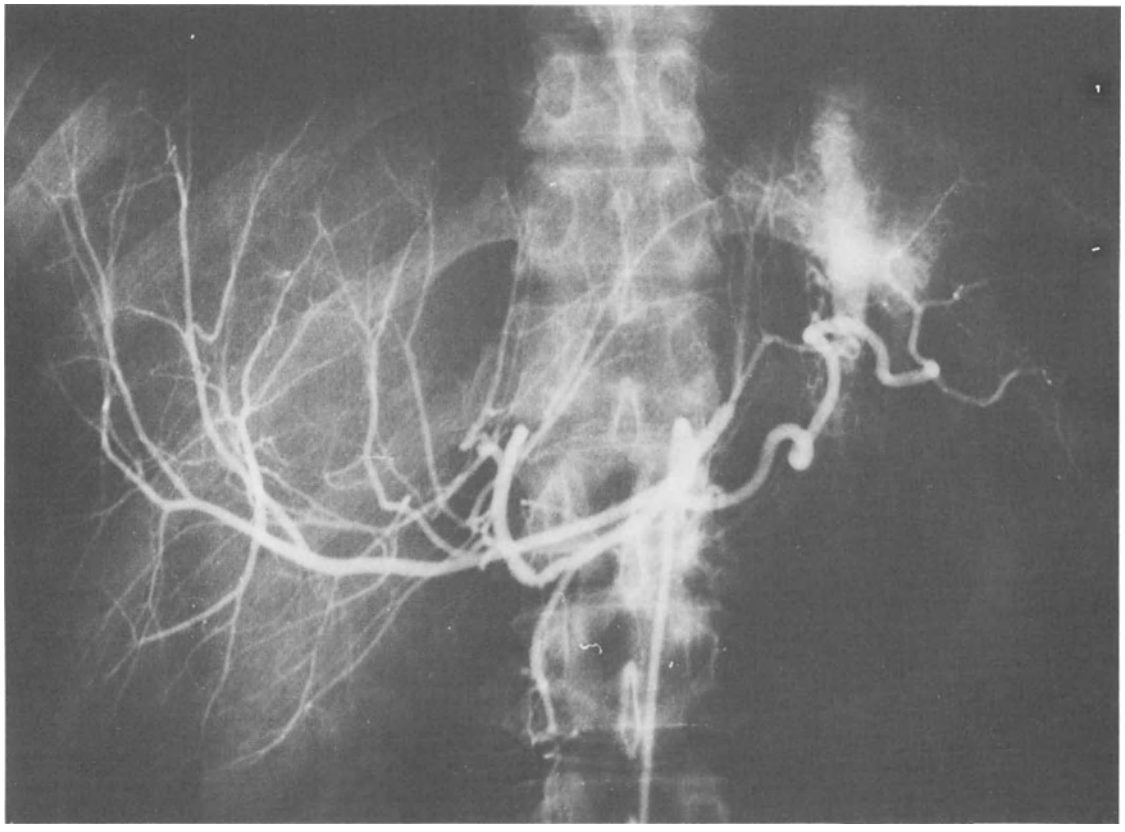


49b

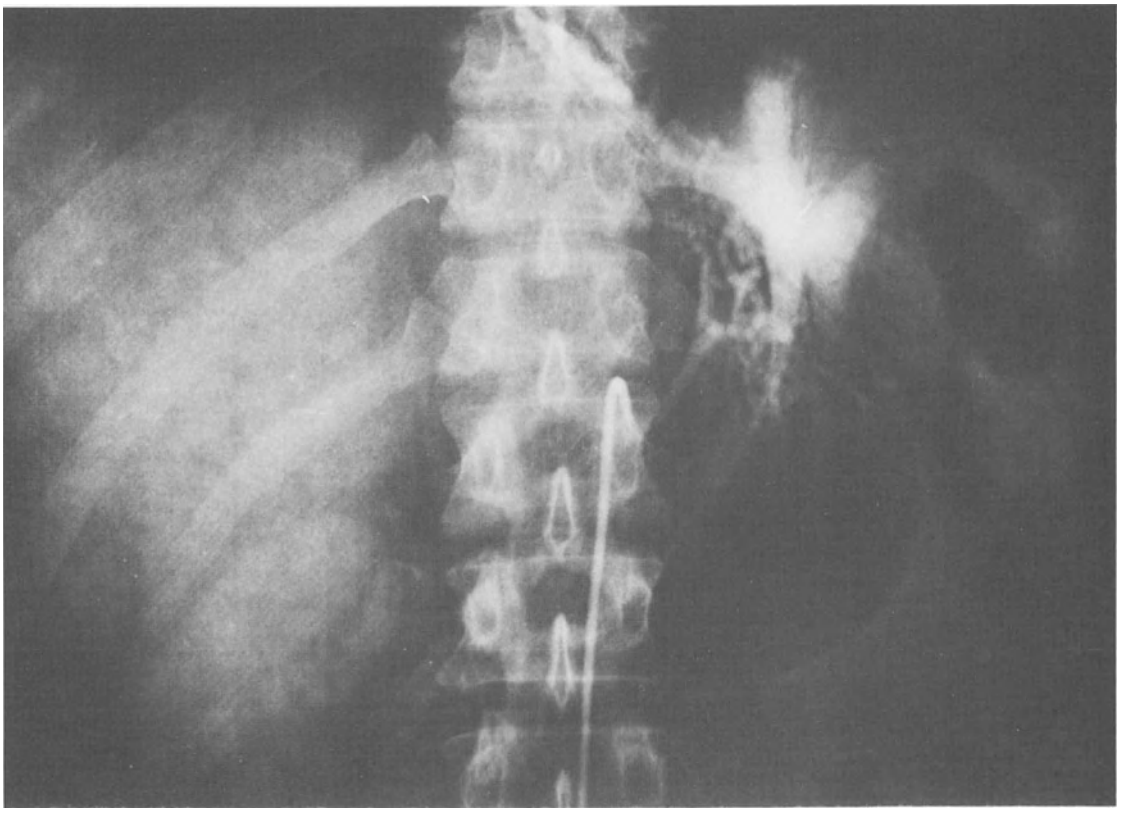
Fig. 49. Alcoholic cirrhosis with a complete obstruction to the portal flow associated with minimal portal hypertension in a 58-year-old female (21–3–79).

49a: Very dilated hepatic artery suggesting a markedly reduced portal flow associated with a normal sized spleen (13 cm) with early and dense splenography. This normal splenic pattern signifies the portal pressure is normal or only minimally and recently increased.

49b: Venous phase of S.M.A.: Completely hepatofugal mesenteric venous flow draining towards a high flow rate spontaneous spleno-renal anastomosis and inferior vena cava through the widened left gastric vein and splenic vein.
Functional obstruction of the portal vein (which was faintly opacified on the venous phase of coeliac arteriography). Despite the completely interrupted hepatopetal portal flow the portal pressure was only slightly elevated: 25 cm saline.
6 months earlier a first hemorrhage was thought to be due to peptic disease and this severe cirrhosis was unrecognized.

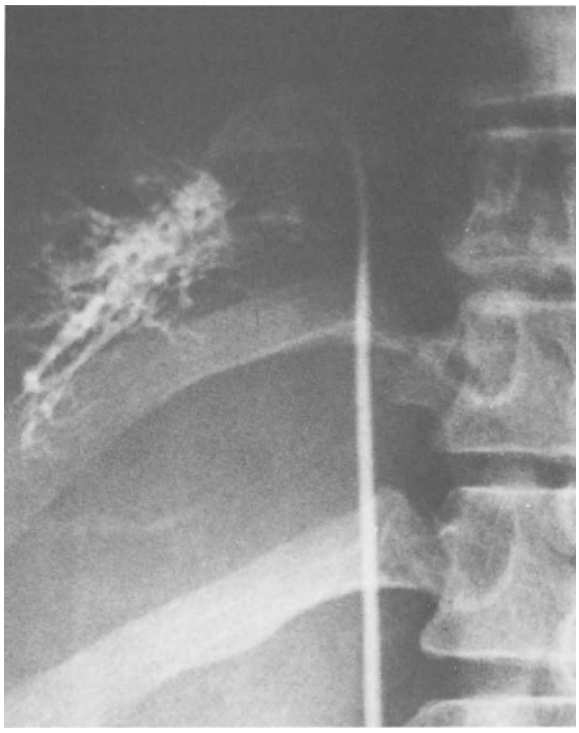


50a



50b

Fig. 50. For legend see next page.



50c

50c: Hepatic venography (8–12–76): obstruction of the hepatic veins with the characteristic spider's web pattern.

←

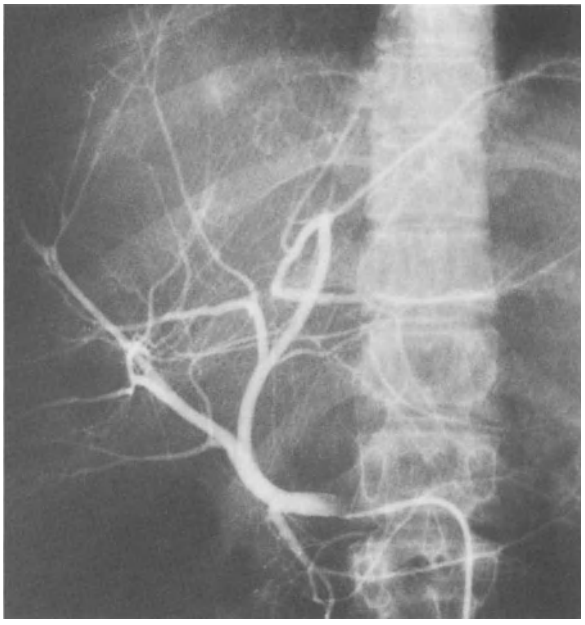
Fig. 50. Budd-Chiari syndrome in a 36-year-old male (16–11–76). Hepatomegaly, ascitis, liver function tests anomalies. Portal pressure: 32 cm saline.

50a: Hepatomegaly with marked enlargement of the left lobe resulting in downward displacement of the hepatic artery. Intrahepatic arterial branches are thin, stretched and regularly spread apart one from the other.

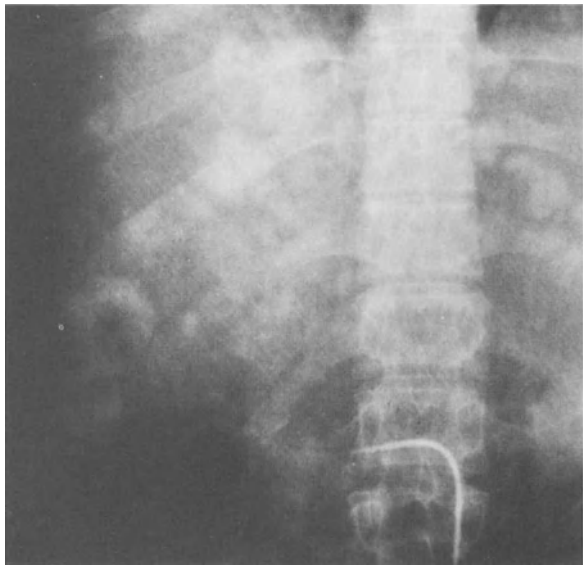
Normal sized spleen with extremely thinned intrasplenic arterial branches (low flow rate spleen pattern).

50b: Venous phase of coeliac arteriography: very faint hepatography and splenography without visualization of the splenic vein.

Owing to the markedly reduced hepatic and splenic blood flows a greater amount of contrast medium enters the gastric arteries resulting in dense opacification of gastric veins and esophageal varices.



51a



51b

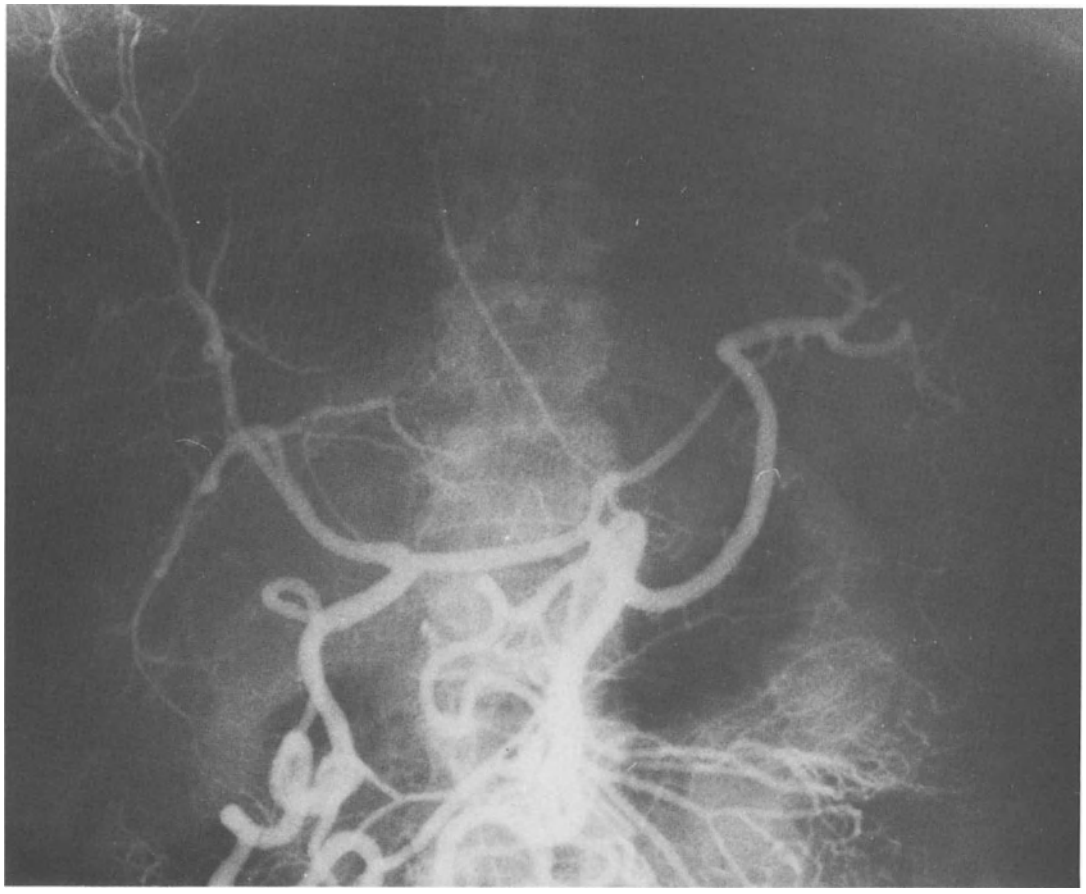
Fig. 51. Budd-Chiari syndrome known for 2 years in a 25-year-old male (82–11–82).

Complete obstruction of the hepatic veins with a patent inferior vena cava confirmed by hepatic venography on 30–9–80.

51a: Pronounced hepatomegaly with very large left lobe. The intrahepatic arterial branches are stretched, thinned and regularly spread apart from each other: Typical pattern of the Budd-Chiari syndrome.

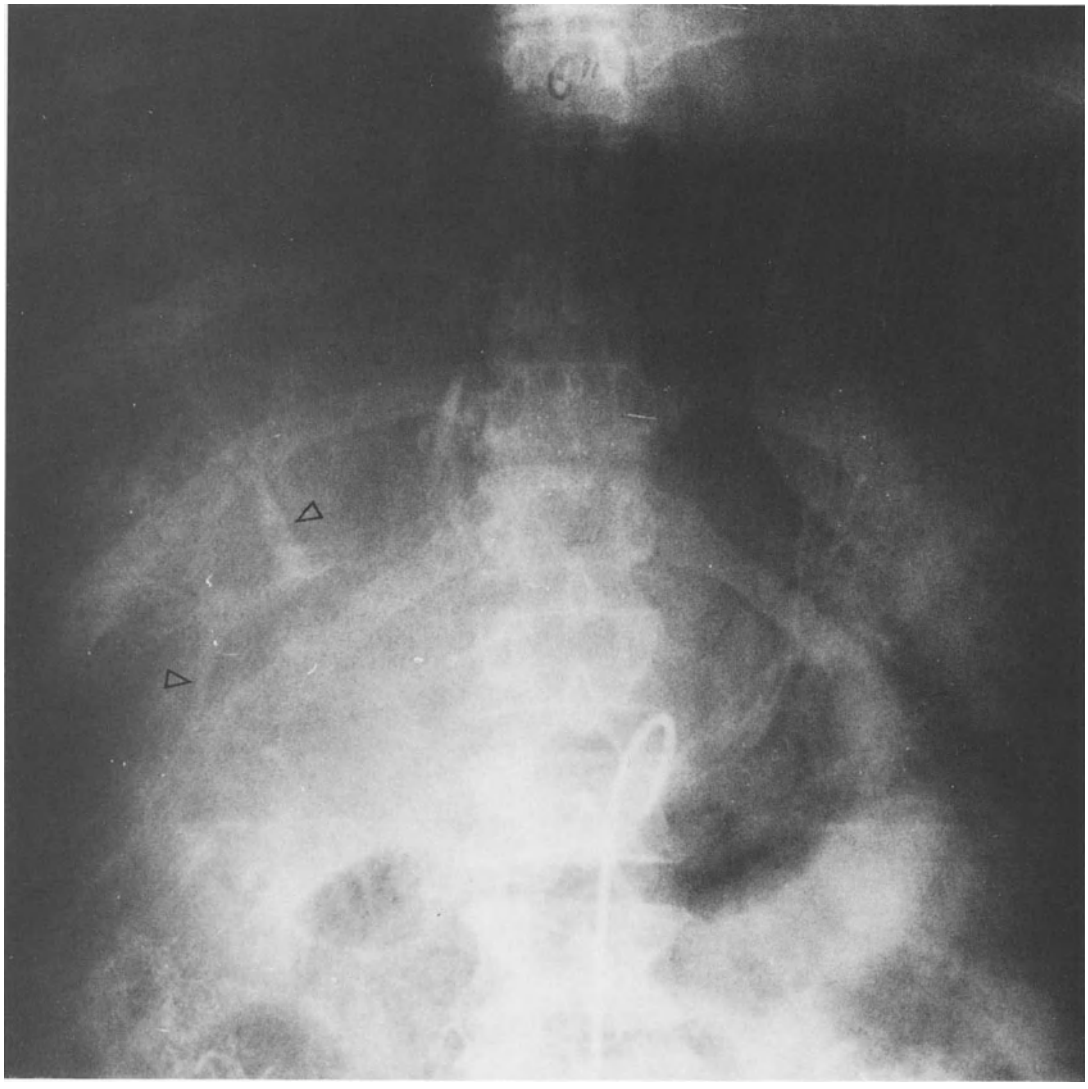
51b: Faint and non-homogeneous arterial hepatography with a few dense patchy areas which should not be confused with metastasis.

Fig. 52. Budd-Chiari syndrome due to primary hepatic vein thrombosis in a 15-year-old female. Same patient as in Fig. 10. Venous phase of coeliac arteriography (1–2–1971). Very faint and heterogeneous arterial hepatography (the splenic vein was not opacified) with a thin stripe of contrast medium outlining the liver margin (↑).



53a

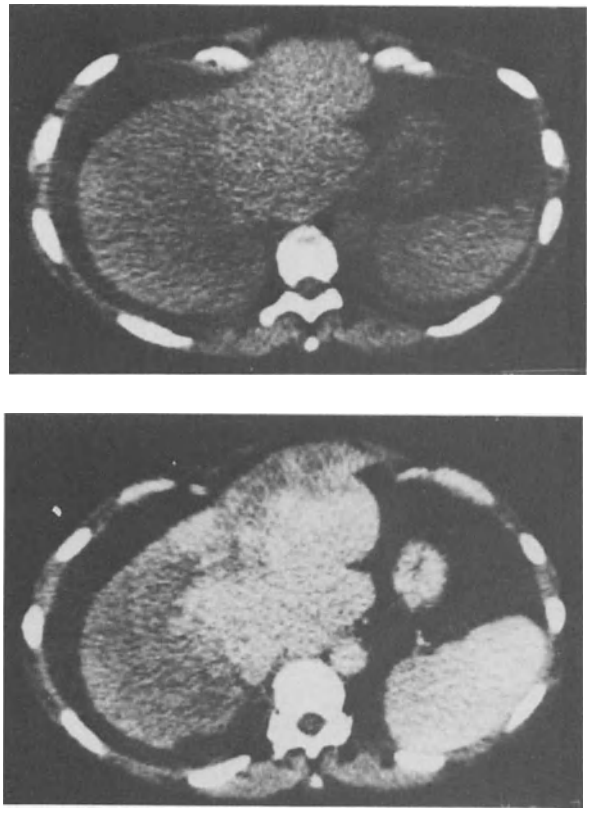
Fig. 53. Budd Chiari syndrome in a 53-year-old female known to have polycythemia for 6 months (12–9–80).
53a: Minimal splenomegaly (15 cm) with the 'low flow rate pattern of the spleen.'
Hepatomegaly with stretched and thinned intrahepatic arterial branches and downward displacement of hepatic artery by the enlarged left lobe.



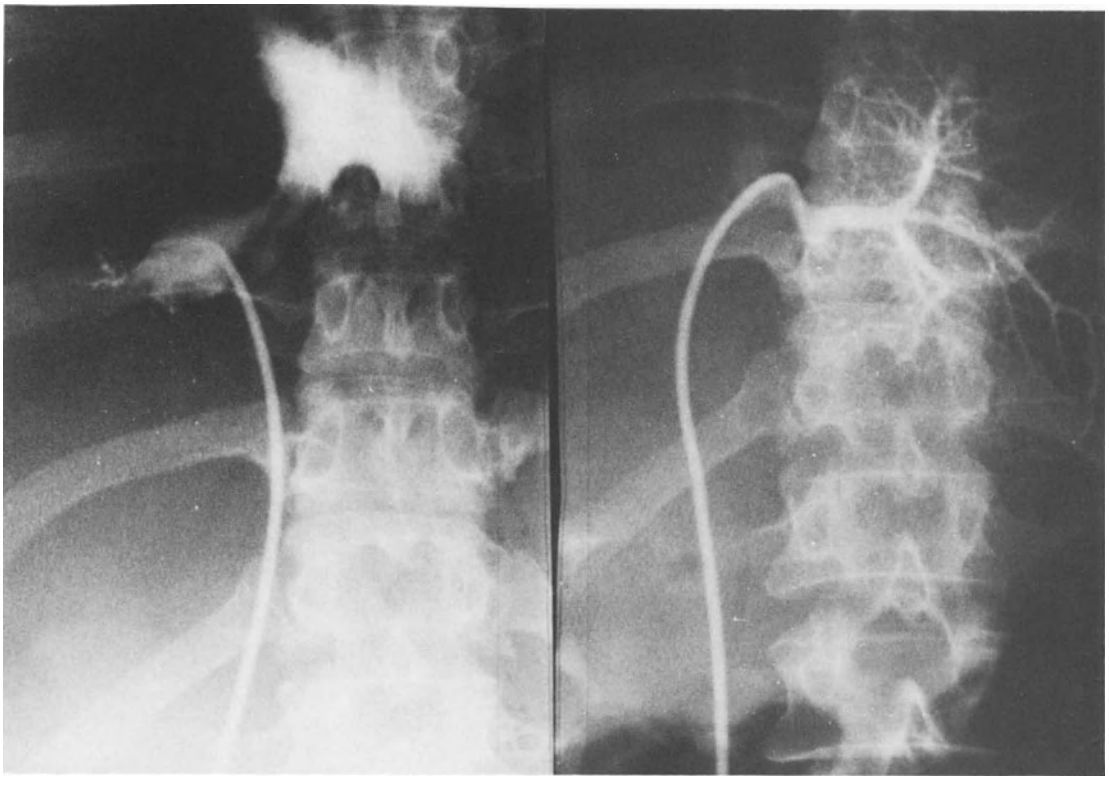
53b

53b: 6 sec later: retrograde filling of some intrahepatic portal branches accounting for the reversal of intrahepatic portal flow (△).

53c: Angio C.T. (12–9–80): Before contrast medium injection a slight hypodensity of the right lobe is visualized as compared to the hypertrophic left lobe and caudate lobe. 30 sec after bolus injection of the contrast medium, normal hepatography in seen only in the caudate lobe and a part of the left lobe whereas the right lobe is not opacified.

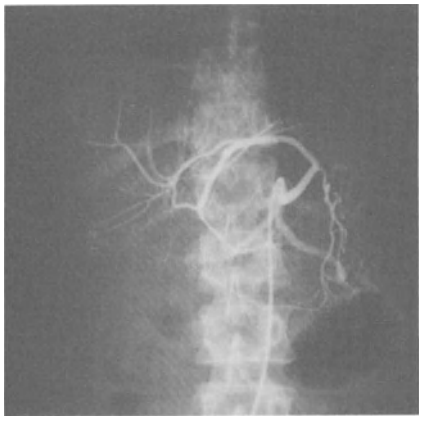


53c

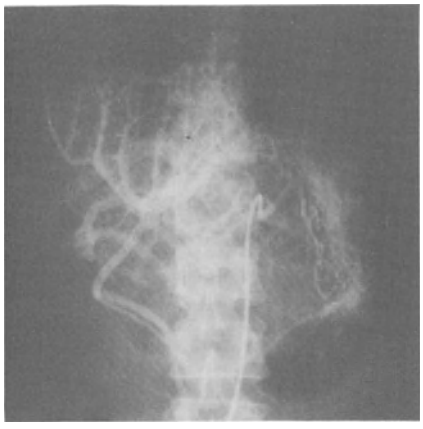


53d

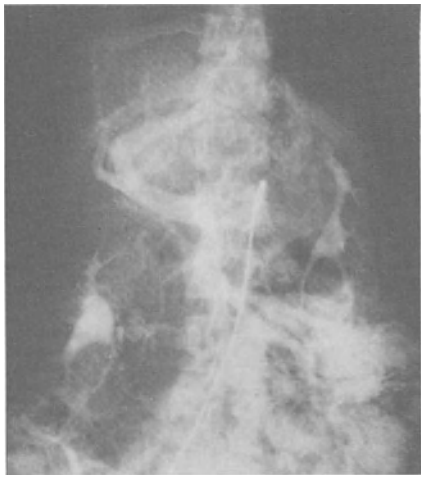
53d: Hepatic venography (15–9–80): The right hepatic vein is completely occluded while partial patency of the left hepatic vein is evidenced.



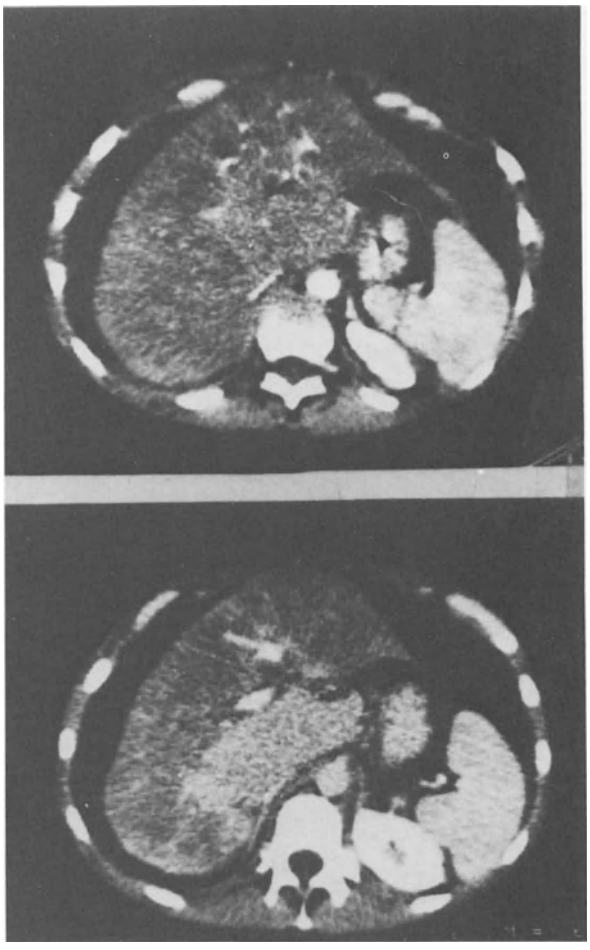
54a



54b



54c



54d

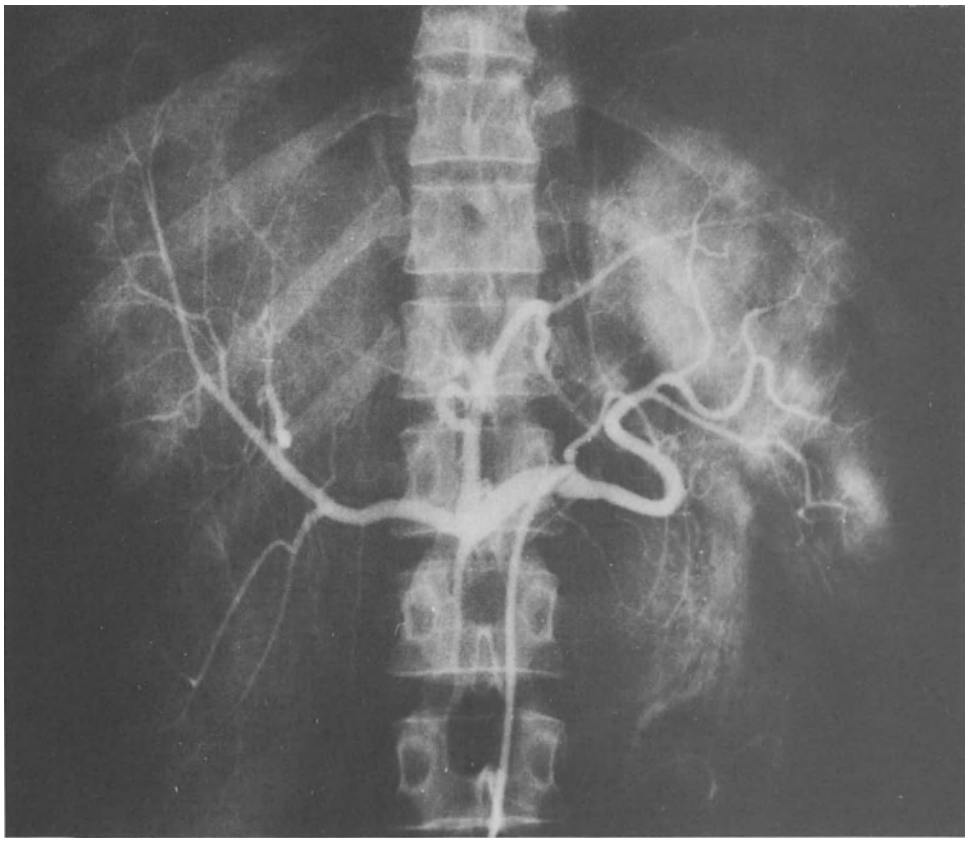
Fig. 54. Budd-Chiari syndrome due to polycythemia in a 40-year-old female.
Patient referred for hepatomegaly and ascitis (18–3–80).

54a: Selective opacification of the left hepatic artery: enlargement of the left lobe with stretched and thinned intrahepatic arterial branches.

54b: 3 sec later, massive arterio-portal shunting within the left lobe with hepatofugal flow through a para-umbilical vein.

54c: Venous phase of S.M.A.: Very poor flow through the narrowed right portal vein branch. The left portal branch is wider and result in a dense opacification only of the caudate lobe.

54d: Angio C.T. (21–3–80) 10 sec after bolus injection of contrast medium the arterial phase demonstrates an early filling of the intrahepatic portal branches within the left lobe, evidencing the arterio-portal shunting.
30 sec later, the venous phase shows that portal hepatography is restricted to the caudate lobe. Owing to this, right and left hepatic veins should be completely occluded as it was confirmed by hepatic venography.



55a

55b

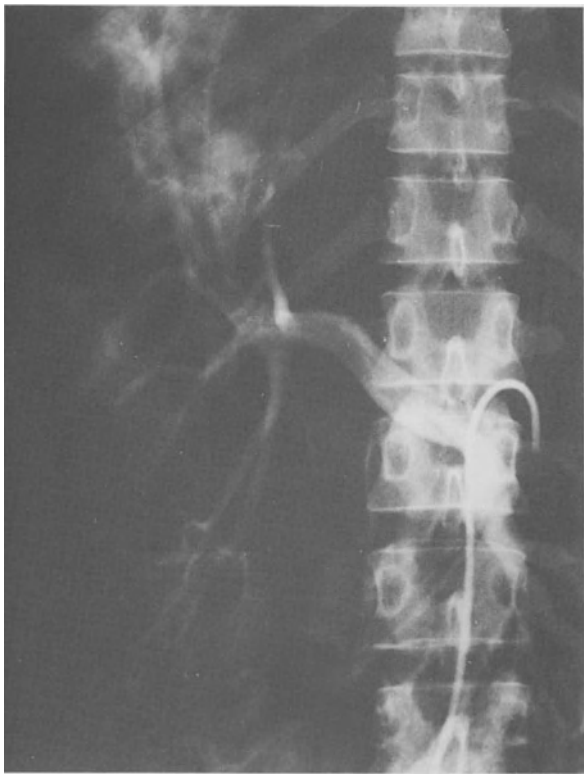
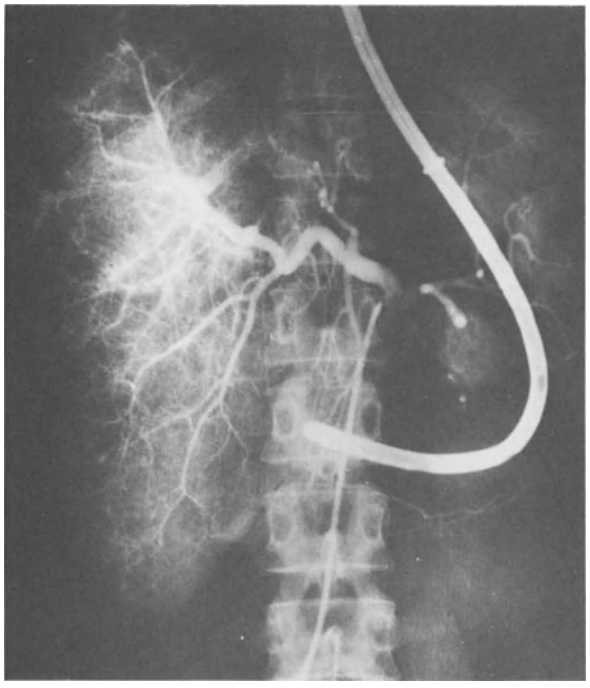


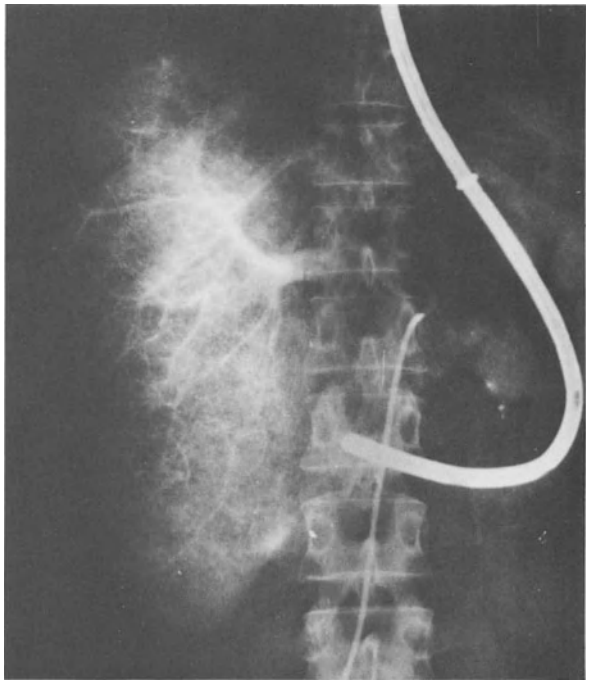
Fig. 55. Budd-Chiari syndrome in a 39-year-old female treated for breast carcinoma (22–10–76).

55a: Moderate splenomegaly (18 cm) with thin intrasplenic arterial branches and faint opacification of the splenic vein on the late films suggesting a relatively severe portal hypertension. Marked and diffuse enlargement of the liver with very thinned and stretched intrahepatic arterial branches.

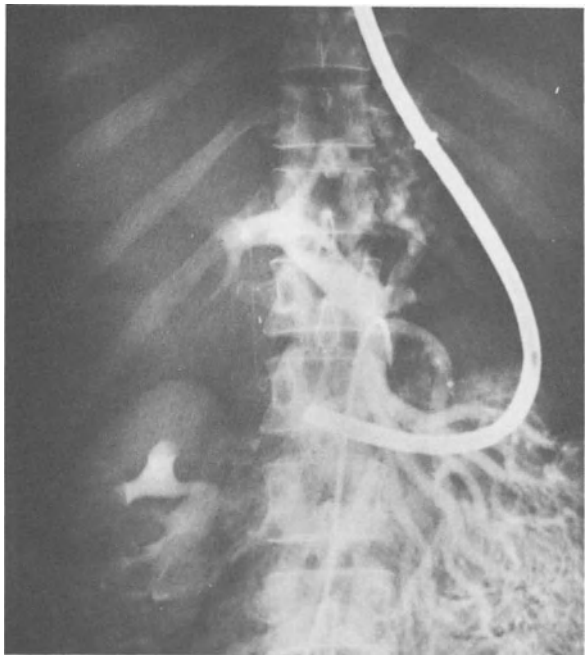
55b: Venous phase of S.M.A.: Poor and non-homogeneous portal inflow, some intrahepatic portal branches resulting in patchy opacification of the liver parenchyma, while most others are not or only faintly visualized.



55c



55d

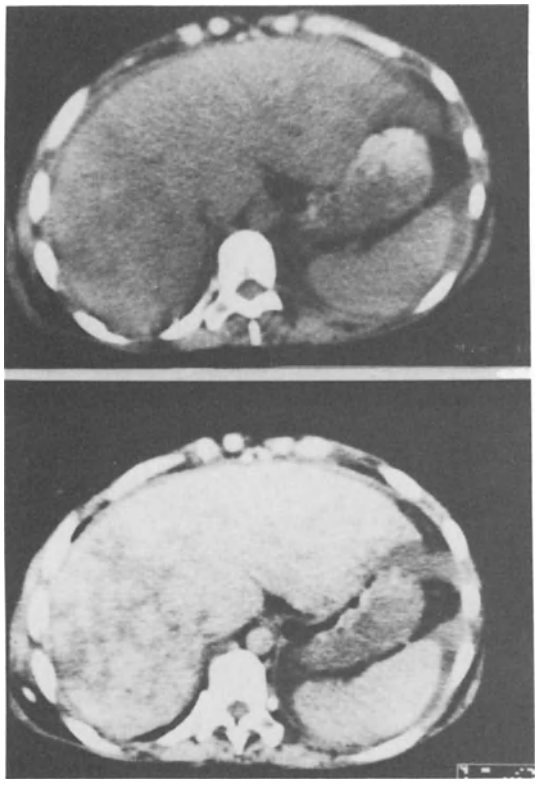


55e

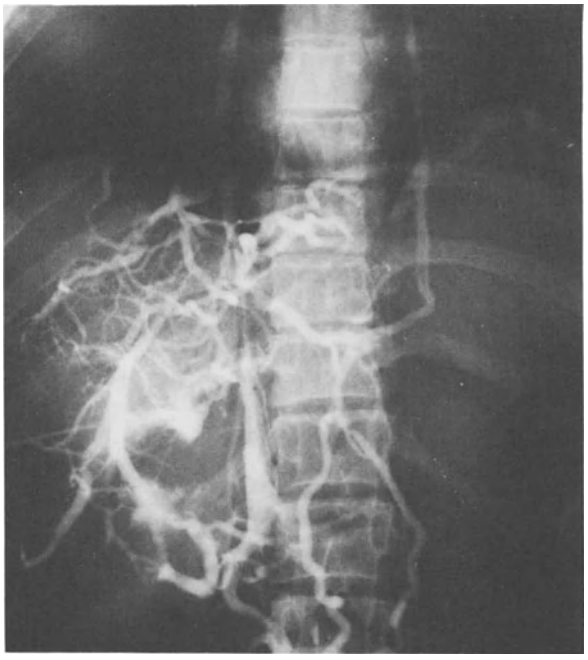
55c: 21 months later, arteriography is performed again after a massive bleeding from esophageal varices (10–7–78). Owing to the development of some porto-systemic collateral pathways, the hepatic artery dilated.

55d: Increased arterial flow resulted in massive arterioportal shunting 4 sec later.

55e: Functional obstruction of the portal vein resulting from the reversed intrahepatic portal flow.



56a

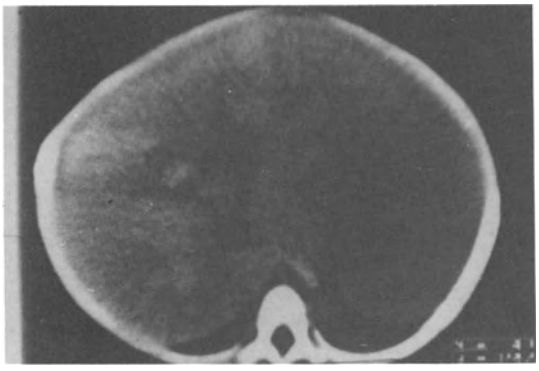


56b

Fig. 56. Budd-Chiari syndrome due to complete thrombosis of hepatic veins and inferior vena Cava in a 34-year-old female (13–12–83).

56a: Angio C.T.: Minimal ascitis and slightly heterogeneous liver before injection. After bolus injection the venous phase shows the distribution of the contrast medium within the right liver lobe to be non-homogeneous.

56b: Percutaneous hepatography: Percutaneous puncture of an intrahepatic hepatic vein demonstrates a dense collateral venous network mainly through the diaphragmatic and internal mammary veins, thus explaining the relatively good portal hepatography documented by angio C.T. Obstruction of the inferior vena cava also visualized.



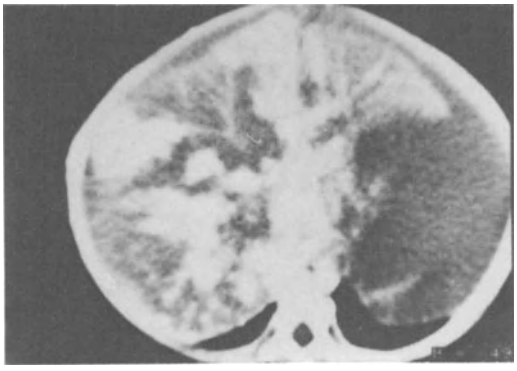


Fig. 57. Budd-Chiari syndrome due to complete thrombosis of the hepatic veins with a patent inferior vena cava in a 4-year-old child.

C.T. scan (29–4–82) shows non-homogeneous pattern of the liver and ascitis.

After intravenous injection of contrast medium a markedly heterogeneous hepatography is evidenced.

A typical pattern of the Budd-Chiari syndrome was also demonstrated by arterio portography (23–4–82).

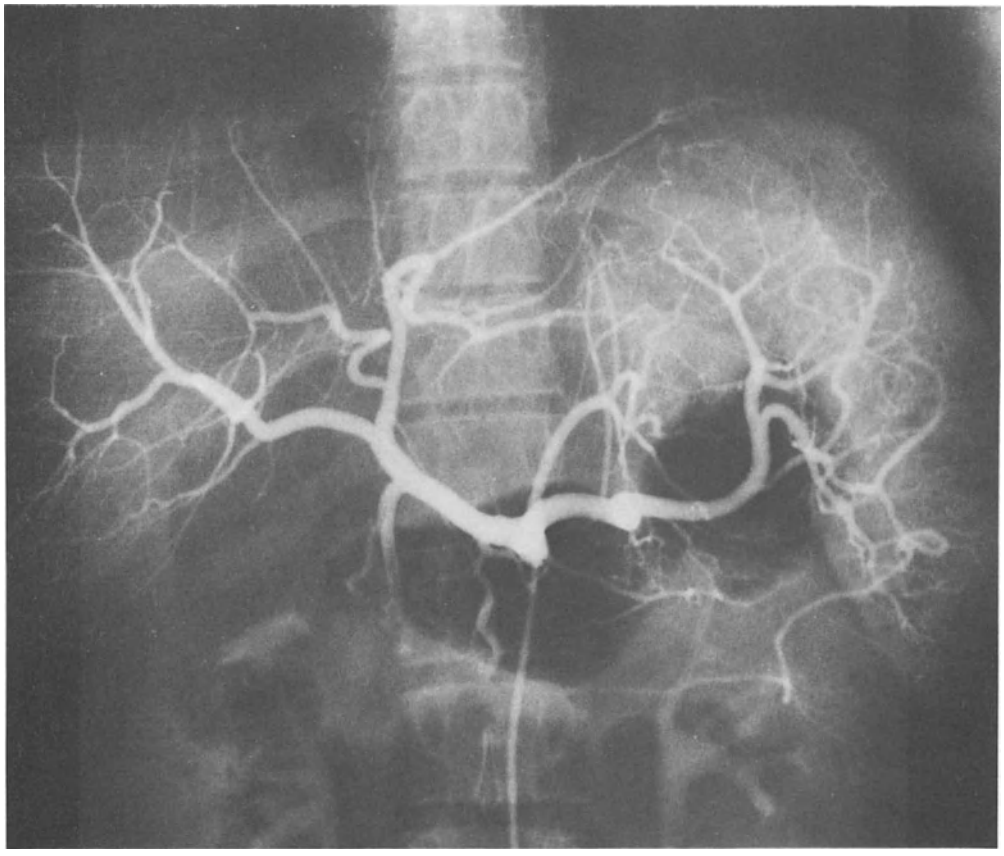
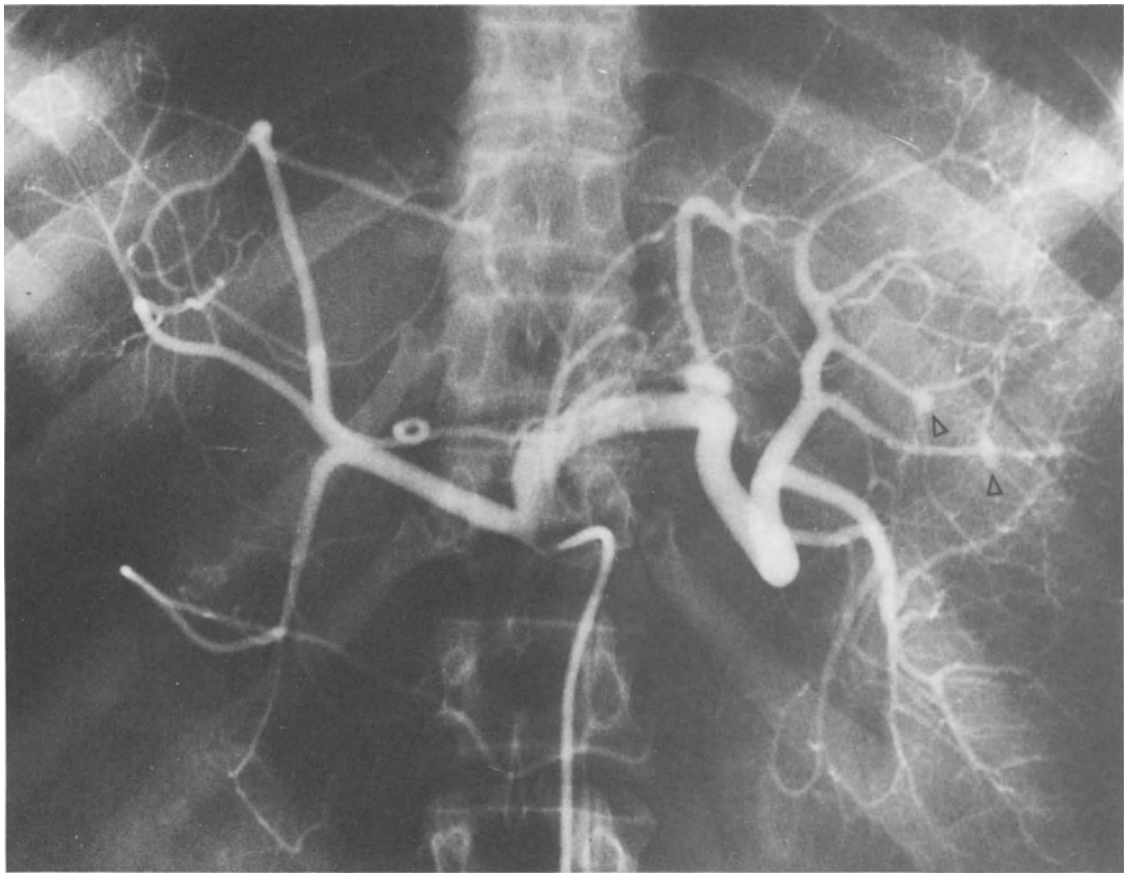


Fig. 58. Portal hypertension resulting from constrictive pericarditis (Pick's Syndrome) in a 17-year-old male referred for ascitis from unknown origin (13–10–81).

- Coeliac arteriography shows the typical pattern of Budd-Chiari syndrome, i.e. an enlarged liver with thinned and stretched intrahepatic arterial branches which are regularly spread apart from each other.
- However, a greater splenomegaly (19 cm) without the usual 'low flow rate pattern' of the spleen signifies the less severe portal hypertension.



59a

59b

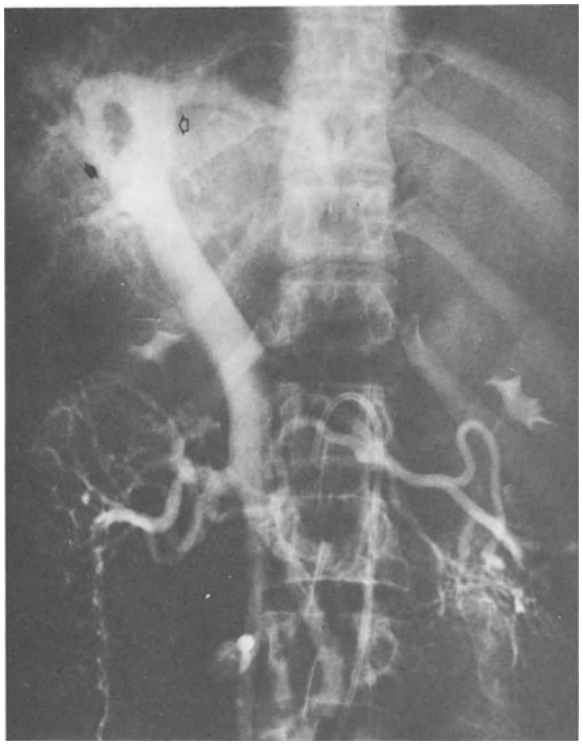


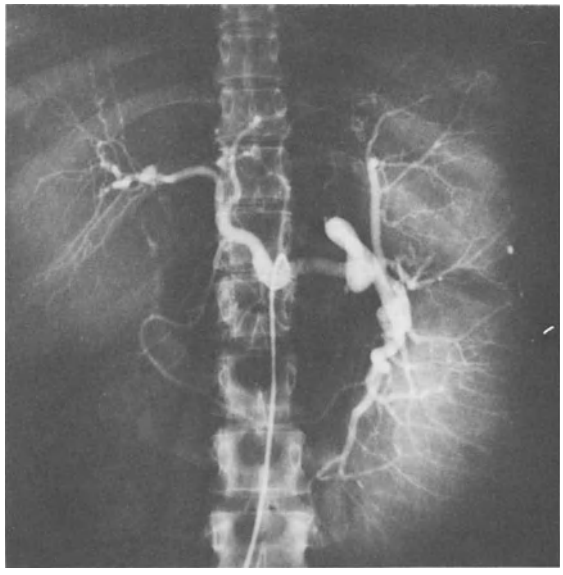
Fig. 59. Atropho-hypertrophic cirrhosis from unknown origin in a 24-year-old male originating from North Africa. Patient referred for variceal hemorrhage (7–7–77).

59a: Markedly enlarged spleen (20 cm) with two small intrasplenic aneurysms (△) associated with an atrophy of the right lobe of the liver.

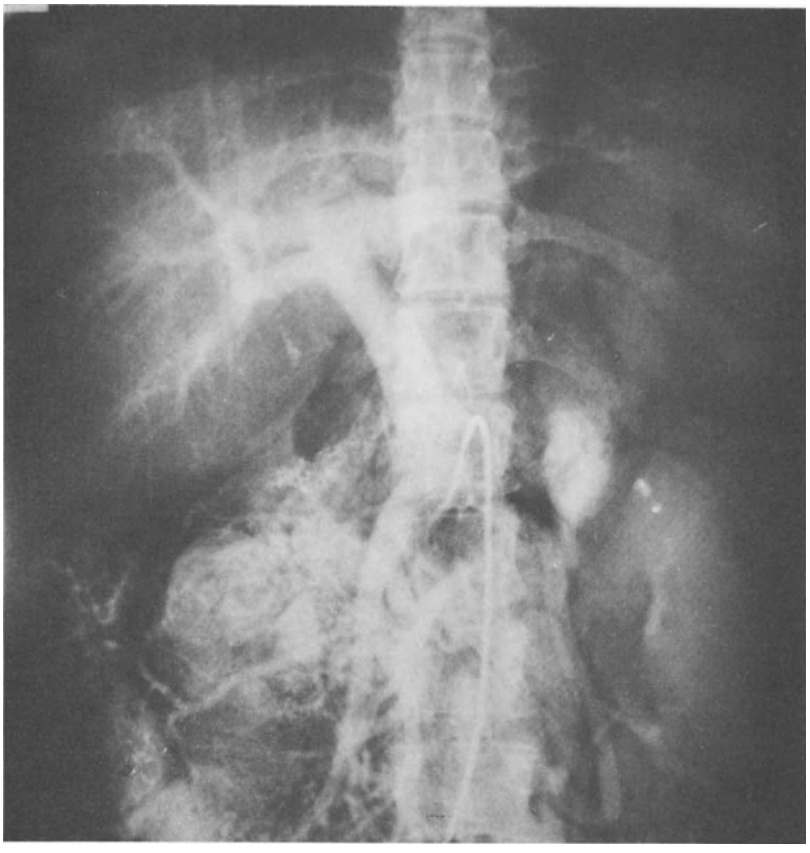
The intrahepatic arterial branches have a normal caliber.

59b: Good opacification of the portal vein and its intra-hepatic branches which signifies a relatively good hepatopetal flow whereas a faint visualization of the inferior mesenteric vein accounts for a very poor hepatofugal flow.

Atrophy of the right lobe of the liver results in narrowing of the right portal branch (↑) as compared to the dilated (⌓) left one which indicates that the left liver lobe is hypertrophied.



60a

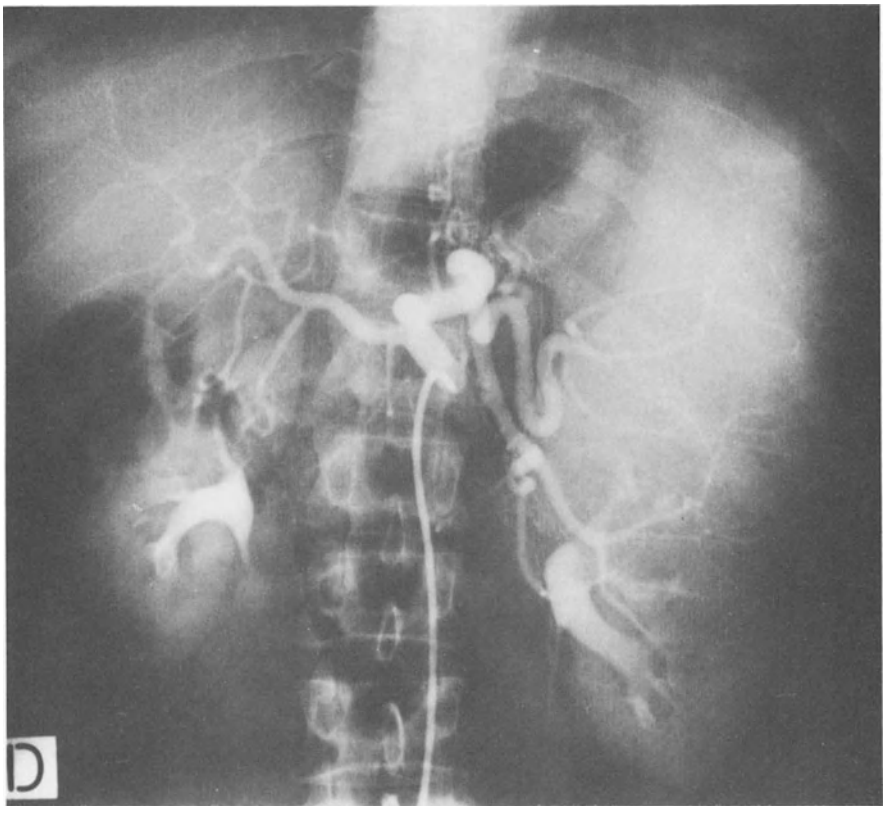


60b

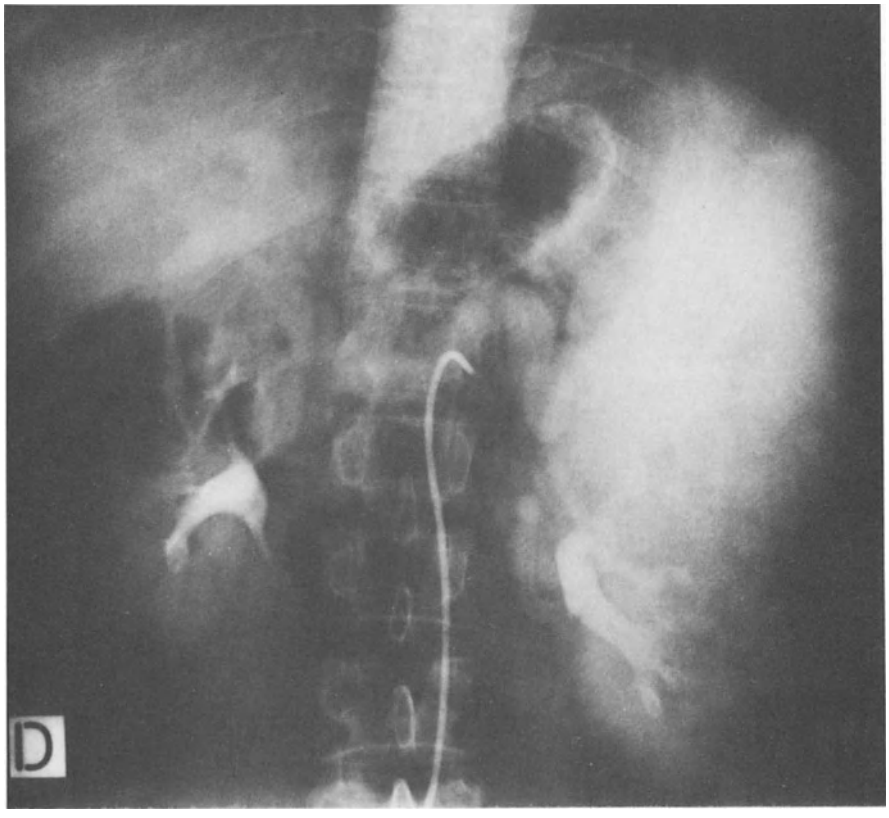
Fig. 60. Cirrhosis probably of the post-hepatitis type in a 32-year-old male.
- Patient referred for splenomegaly, variceal bleeding and jaundice (19–9–78).
- Two previous episodes of virus hepatitis at the age of 6 years and 30 years.
- Corrected sinusoïdal pressure: 37 cm saline.

60a: - Huge splenomegaly (28 cm) with dilated and lengthened splenic artery suggesting a slowly progressive and long-standing portal hypertension.
- Diffuse and moderate hepatomegaly without dilatation of the hepatic artery accounting for a minimal obstruction.

60b: - Venous phase of S.M.A.: Normal or near normal opacification of the intrahepatic portal branches which signifies the good hepatopetal portal flow, whereas only a faint opacification of the inferior mesenteric vein is seen.

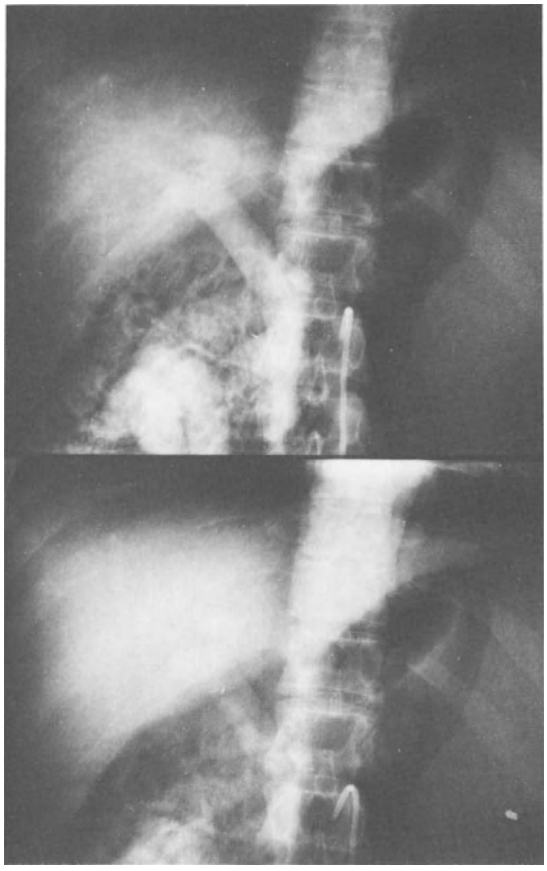


61a



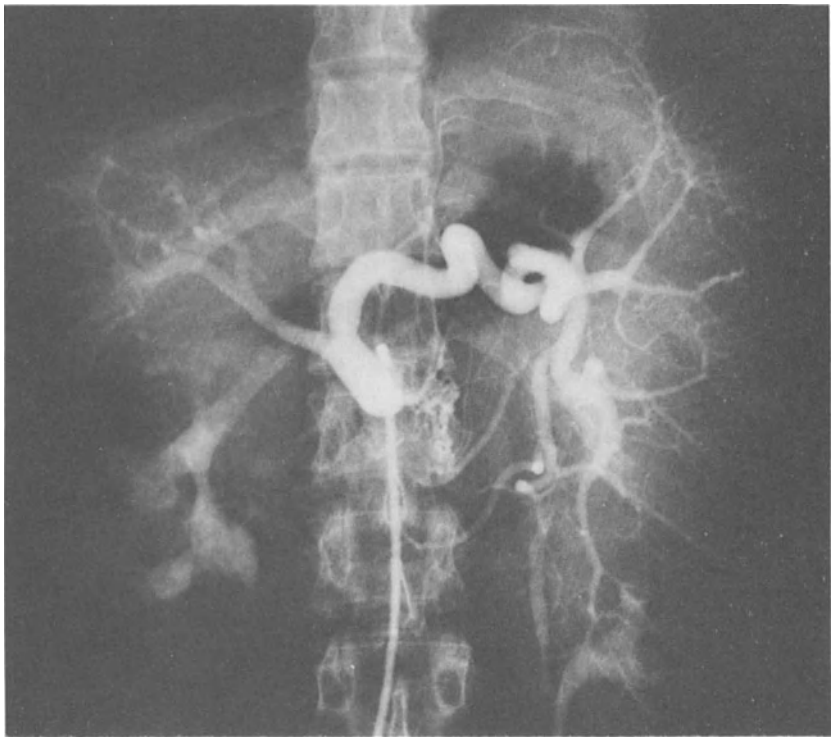
61b

Fig. 61. For legend see next page.



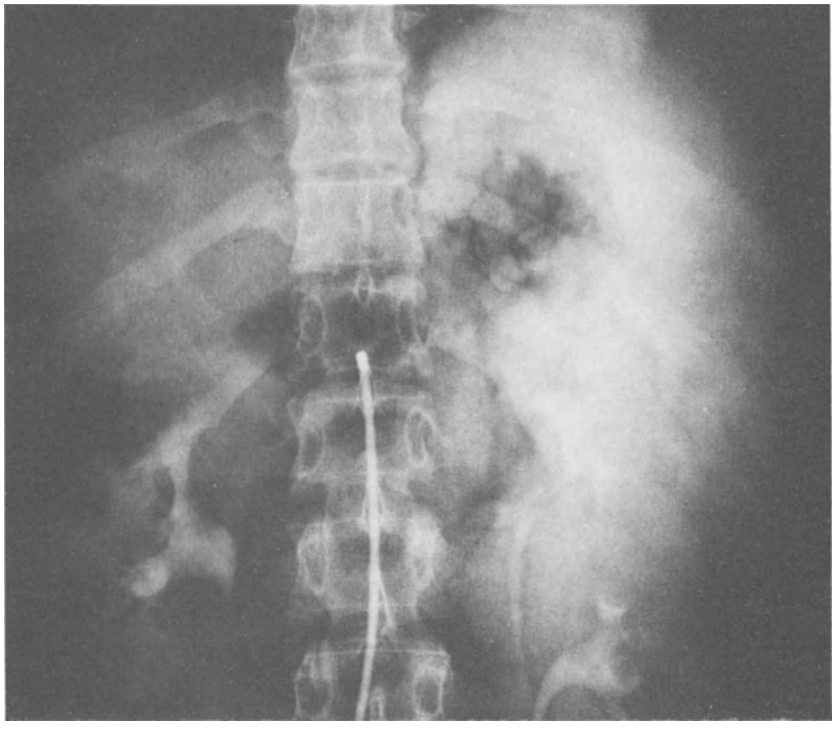
61c

Fig. 61. Post-hepatitis cirrhosis (or liver fibrosis) in a 38-year-old male.
– Patient referred for splenomegaly, known since 1961, mild jaundice and asthenia (6–10–75)
– Two previous episodes of virus hepatitis in 1958 and 1962.
61a: – Huge splenomegaly (28 cm) with dilated and lengthened splenic artery.
– Normal pattern of the liver and normal sized hepatic artery.
61b: – Dense and homogeneous splenography with dilated and faintly opacified splenic vein.
61c: Venous phase of S.M.A.:
Normal portal vein resulting in dense portal hepatography which signifies the normal or near normal hepatopetal portal flow. No hepatofugal flow is visualized.
– Splenectomy is performed (29–10–75).
– portal pressure 22 cm saline lowered to 18 cm saline after ligation of splenic artery.
– fibrocongestive spleen weighing 1,400 g.
– histological pattern suggesting a fibro-muscular dysplasia of splenic artery.
– liver biopsy: periportal fibrosis.



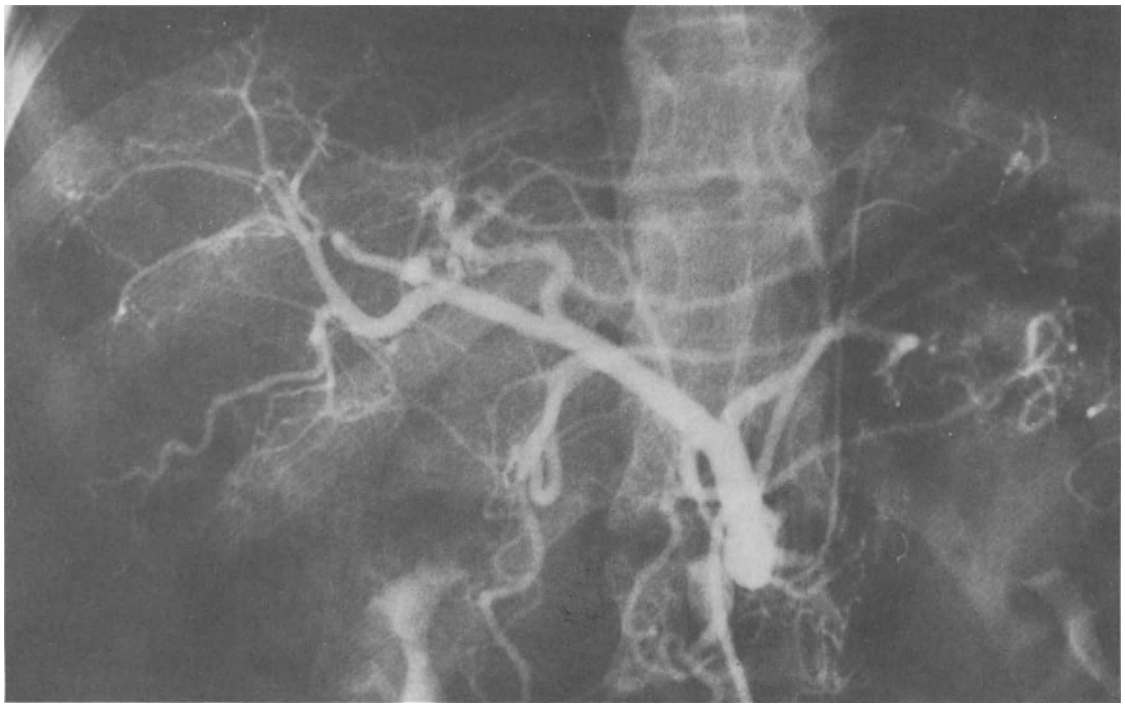
62a

Fig. 62. Cirrhosis from unknown origin in a 29-year-old female.
Patient found to have splenomegaly with hypersplenism during pregnancy (7–12–73).
62a: Huge splenomegaly (27 cm) with dilated and lengthened splenic artery and normal liver.



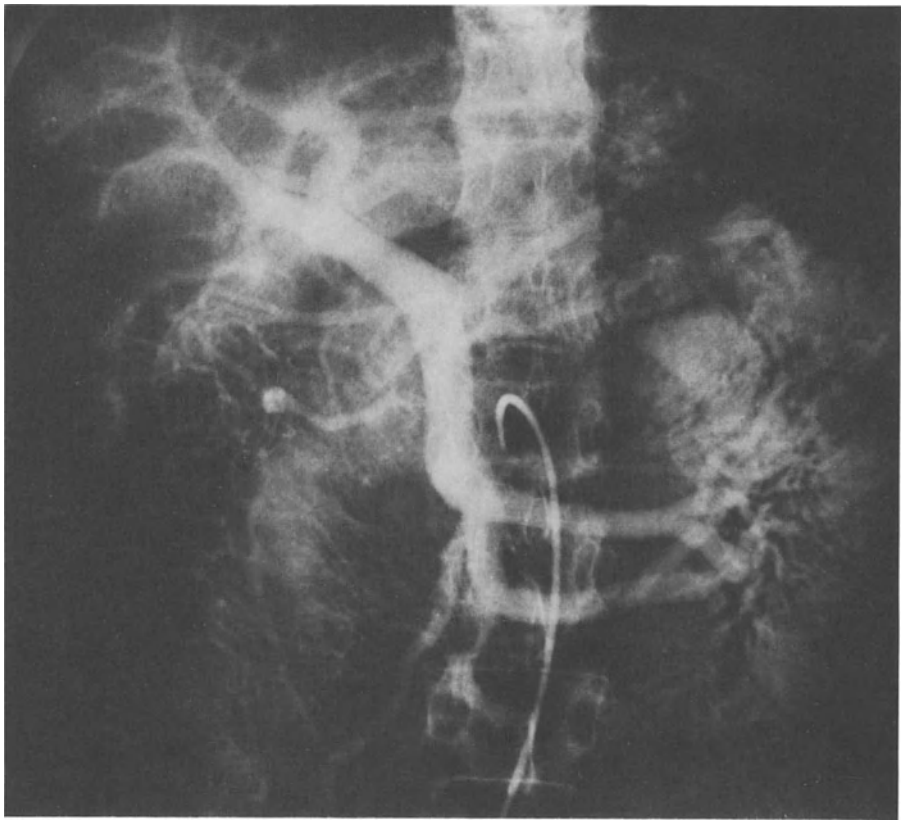
62b

62b: Dense and homogeneous splenography with very poor opacification of the splenic vein and faint visualization of dilated and tortuous short gastric veins indicating a splenic blood stasis.
– splenectomy performed in 8/75. Liver biopsy: mild periportal fibrosis.
– esophageal varices bleeding 4 years later.



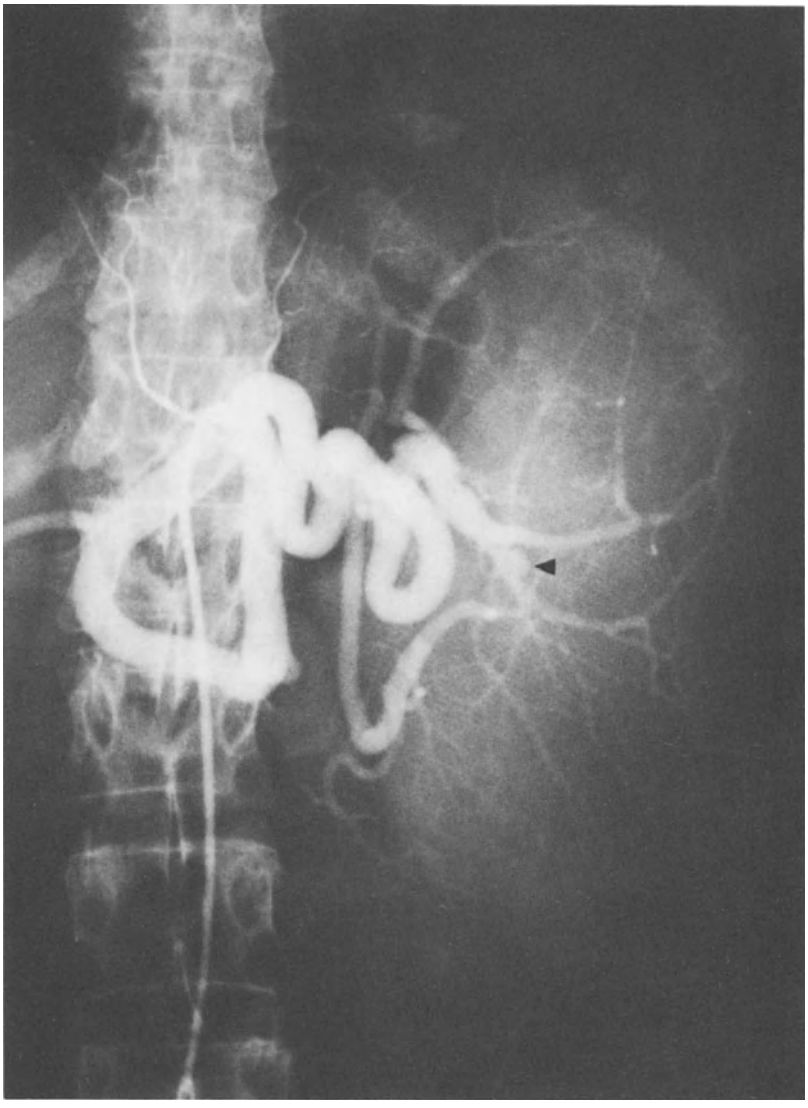
62c

62c: Coeliac arteriography (9–10–79).
Atrophy of the liver and minimal dilatation of the hepatic artery.



62d

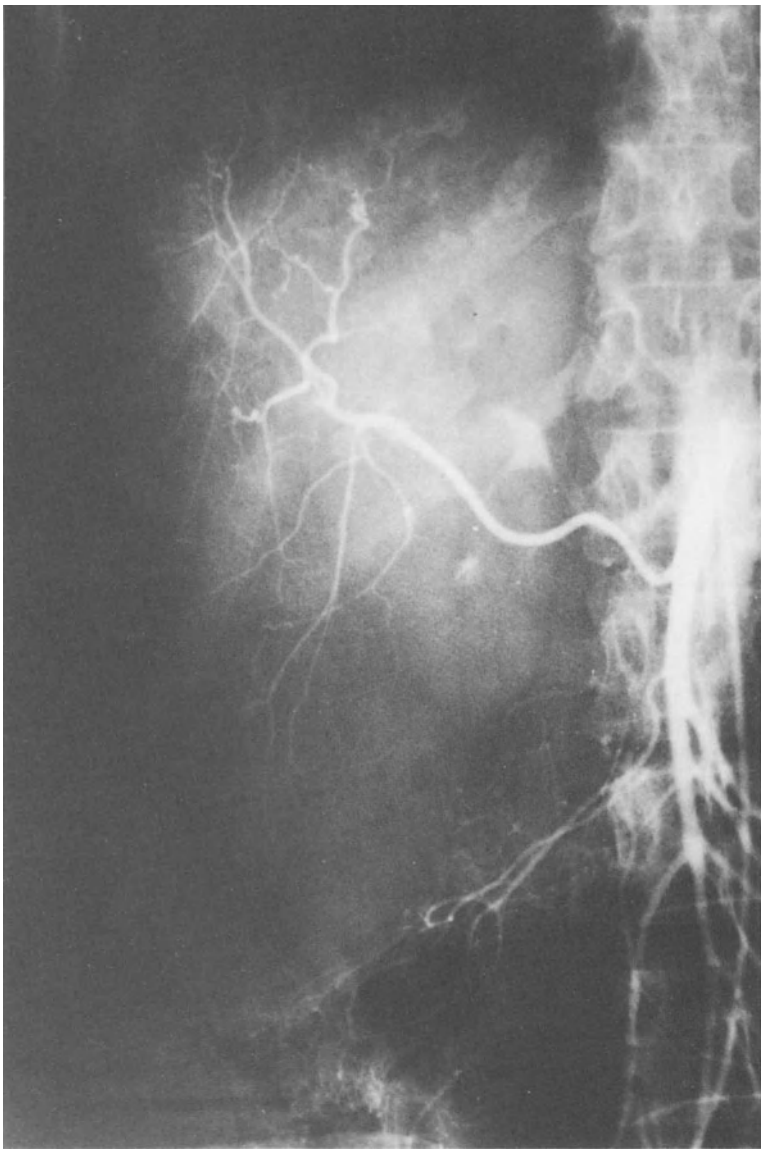
62d: Venous phase of S.M.A.: relatively good opacification of slightly narrowed intrahepatic portal branches with retrograde flow in the left gastric vein which signifies the moderately decreased portal inflow. Liver biopsy: cirrhosis.



63a

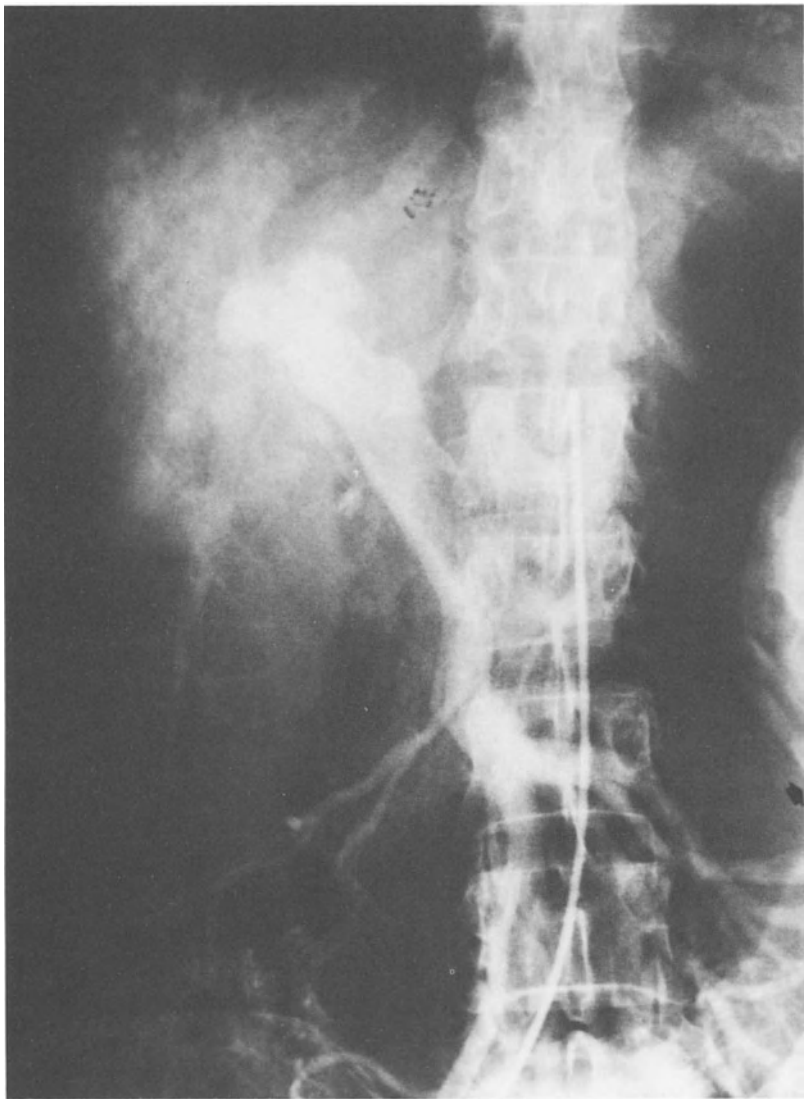
Fig. 63. Presinusoïdal intrahepatic obstruction revealed by splenomegaly with hypersplenism in a 62-year-old female from Spain (10–5–84).

63a: Very large spleen (23 cm) with widened and lengthened splenic artery and a small intrasplenic aneurysm (▲): typical pattern for a long-standing and slowly progressive portal hypertension.



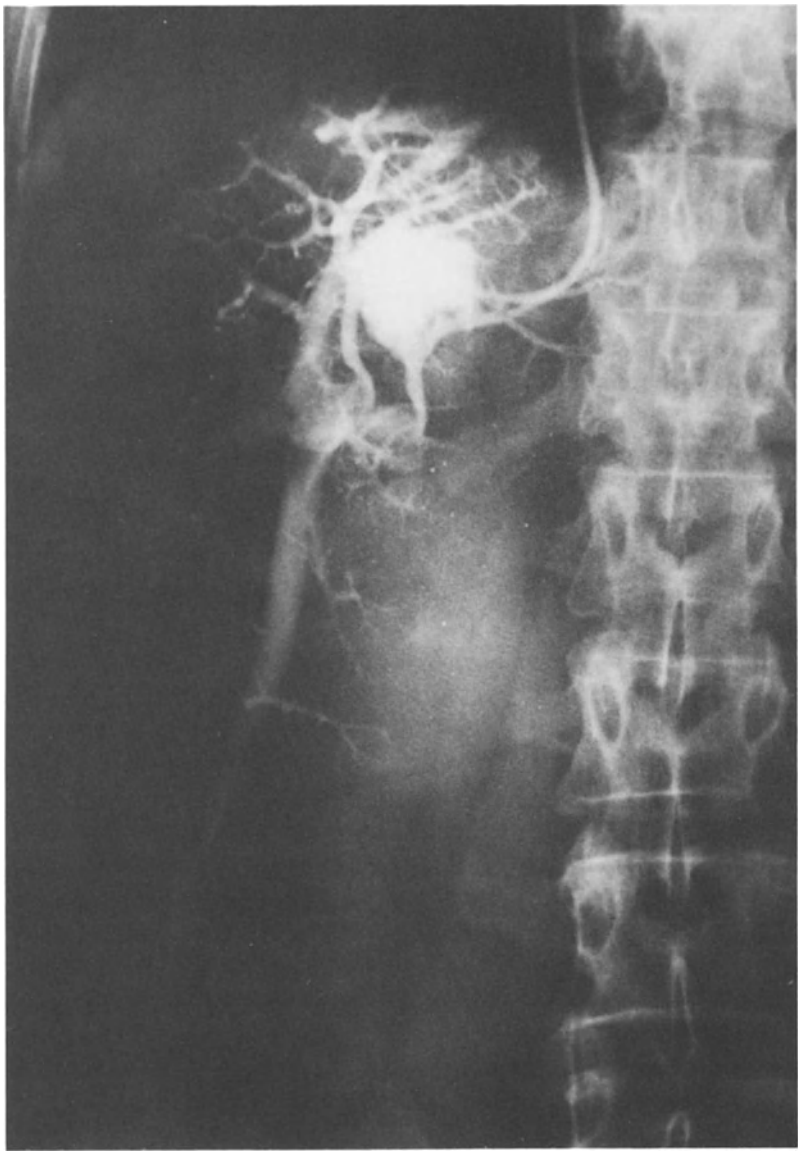
63b

63b: Thin right hepatic artery with slightly tortuous small intrahepatic branches in the upper part of the right lobe suggesting segmental atrophy of the liver. The upward displacement of the right kidney also accounts for atrophy of the right lobe of the liver.



63c

63c: Venous phase of S.M.A.: widened portal vein with moderately decreased portal inflow and minimal reflux in the inferior mesenteric vein (↑).



63d

63d: Wedged hepatic venography (15–5–84): retrograde opacification of the intrahepatic portal veins demonstrating atrophy of the upper right lobe of the liver. The corrected sinusoïdal pressure is normal: 4 mm Hg. Liver biopsy was not conclusive. This angiographic pattern suggests idiopathic portal fibrosis.

6. ARE THERE PORTAL HYPERTENSIONS WITHOUT OBSTRUCTION?

Three essential factors can cause portal hypertension: (a) an increase in circulatory resistances (b) inefficiency of the compensatory collateral circulation, and (c) an increase of the portal vein flow. The most frequent cause, of course, is the increase of the intrahepatic resistances, which leads to a decrease in the hepatopetal portal flow.

An abnormal increase of the portal flow can only aggravate the portal hypertension due to a preexisting obstruction. However, certain authors have hypothesized that an isolated increase of the portal flow without any increase of the circulatory resistance can sometimes be the only cause of portal hypertension. This has caused controversy in the field of portal hypertension because it is difficult in practice on the one hand to measure the portal flow and on the other hand to verify the integrity of the liver; some minimal hepatic lesions can escape the percutaneous biopsy.

This forward pressure portal hypertension has been discussed in two different circumstances, whether the increase in portal flow can be attributed to a very large splenomegaly or to the existence of an arteriovenous fistula.

I. Portal hypertension of splenic origin

The hypothesis that a very large splenomegaly with increased splenic venous flow can cause portal hypertension originates from the description of Banti's disease. It has subsequently been described in the European literature under the name of forward pressure portal hypertension (96) and more recently in the American literature under the name of hyperkinetic or active portal hypertension (97, 98).

At the beginning this concept came from the clinical observation of a portal hypertension associated with a very large splenomegaly and an apparently normal liver. In fact the role of the spleen in this portal hypertension has never been demonstrated because in most cases neither the perfect integrity of the liver nor the increase of the hepatopetal portal flow had been clearly proven.

In most of the reported cases, in fact, the hepatic biopsy usually demonstrates periportal fibrotic lesions. In certain cases where the liver is described as normal it is possible that discrete lesions have escaped the needle biopsy, as has been stressed by certain authors.

In our experience we have frequently observed a discrepancy between the histological and angiographic patterns of the liver. The hepatic biopsy shows a normal or almost normal pattern of the parenchyma while the arterioportography demonstrates a serious atrophic or atropho-hypertrophic transformation. This tends to confirm that the needle biopsy does not always demonstrate these hepatic lesions, especially when they are chronic, long-lasting and stationary lesions.

Finally the fact that splenectomy does not inhibit the slow aggravation of these lesions and the evolution toward hepatic insufficiency, tends to prove that in these patients there exists hepatic lesions (Figs. 17, 62). These are probably unknown at the beginning because they are discrete and progress very slowly.

The existence of arteriovenous shunts in these very large splenomegalies has never been demonstrated. These hypothetical shunts would have caused an increase of the splenic venous flow. Even if we can demonstrate an increase of the splenic venous flow, the origin of the splenomegaly remains completely unknown. However, in practice, a very large splenomegaly essentially results from a minimal and slowly progressive obstruction to hepatopetal portal flow, whatever its cause. This explains why these cases of portal hypertension with very large splenomegaly do correspond to an increase of the intrahepatic circulatory resistances and fall into the classification of idiopathic noncirrhotic portal hypertension.

Viamonte has demonstrated, however, that it is

possible that an increase of the splenic venous flow associated with a very large splenomegaly can play a certain role in the origin of the portal hypertension (97). In fact, even if the splenomegaly is primarily the consequence of an obstruction to hepatopetal portal flow, the increase of the splenic size is not uniquely the result of a passive dilation of the intra-splenic vascular spaces due to stasis. The hyperplasia of splenic parenchyma can also be accompanied by an increase of the blood flow, as is suggested by the simultaneous dilation of the splenic artery and vein.

It is not surprising that a moderate venous stasis promotes this splenic hyperplasia. There are other examples in the human body where a venous stasis facilitates the increase and hypertrophy of the corresponding territory. In particular the Klippel-Trenaunay syndrome is an example of a case where the hypertrophy of a limb seems to be the consequence of the agenesis of the corresponding deep venous trunks.

Even if the increase in the size of the spleen is accompanied by an increase in the circulating blood volume, it is possible that the splenic venous flow remains normal if the circulation is decreased because of stasis. It is equally possible that an increase of the splenic venous flow is associated with a decrease of the mesenteric venous flow so that the hepatopetal portal flow remains unchanged. During these portal hypertensions with a very large splenomegaly we have never observed any angiographic pattern which could evoke an increase in the mesenteric or splenic venous flow. Certainly the poor quality of the venous phase of splenic arteriography can be explained by the dilution of the contrast medium in the spleen. However, the venous phase of the superior mesenteric arteriography always demonstrates poor opacification of the intrahepatic portal branches and an absence of portal hepatography which is in relation to a decrease of the hepatopetal portal flow.

However, Viamonte has demonstrated that if the suppression of the splenic blood input leads to a clear decrease of the portal pressure, then the increase in splenic venous flow may be responsible, at least in part, for the portal hypertension. In fact, in one case the wedged hepatic pressure, which was between 32 and 37 mm of mercury, decreased to 13 to 15 mm of mercury after occlusion of the splenic artery by a balloon catheter. This result seems to prove that an increase of the splenic blood flow is capable of strongly aggravating a minimal portal hypertension due to a discrete increase of intrahepatic resistance.

Therefore, it is possible that these minimal hepatic lesions e.g., a discrete periportal fibrosis, lead to a moderate increase of the portal pressure, which is capable of compensating for the increase of intrahepatic resistance so that the hepatopetal portal flow remains practically normal (15).

If these lesions progress slowly or not at all, they will be clinically latent for a long time and can be revealed late by a very large splenomegaly. At this stage, it is certainly possible that the increase of the circulating splenic volume leads to an increase of the splenic venous flow even if the circulatory speed remains normal. This could explain the so-called hyperkinetic portal hypertension.

II. Portal hypertension and arteriovenous fistulas

A certain number of portal arteriovenous fistulas have been reported as the origin of portal hypertension in patients whose liver was apparently normal. These fistulas are usually congenital or more often of traumatic origin. They are located at the level of the mesenteric, gastric, and splenic vessels, or even within the liver. In these cases the increase of portal flow is strongly suggested by the angiographic pattern as well as by the increase of the B.S.P. clearance, and sometimes proven by measuring the flow.

However, the exact role of these arterioportal fistulas as the cause of a portal hypertension remains debatable because the liver is not always strictly normal histologically. After correction of the fistula, the portal pressure decreases but does not always return to normal (99). Finally, a certain number of these arterioportal fistulas are unaccompanied by portal hypertension even though the portal flow is clearly increased (100) (Fig. 21).

Maillard et al., in reviewing 53 arterioportal fistulas reported in the literature, have noted that 23 cases are accompanied by a portal hypertension but that it is usually in relation with associated lesions. Four of these cases of portal hypertension seem to be solely due to the fistula (100). More

recently, Puglionizi et al., reviewing 179 cases in the literature, have noted the association to portal hypertension in 50% of the cases (101). The portal hypertension existed in 82.9% of the splenic arteriovenous fistulas, in 54,5% of the fistulas involving the hepatic artery, and in 29.4% of the mesenteric fistulas.

These authors add that normally the portal flow uses only a part of the intrahepatic venous bed so that the portal flow can increase significantly (between two and three times) without modification of the portal pressure. Therefore, theoretically a forward pressure portal hypertension can only be the consequence of a very large increase in the hepatopetal portal flow.

In our experience of more than 1000 cases of portal hypertension explored by arteriography, we have observed only three cases of arterioportal fistulas. In one case, multiple and very large congenital mesenteric arteriovenous fistulas in a 30-year-old male were unaccompanied by portal hypertension despite the clear increase of cardiac output. In the other two cases the intra-hepatic arterioportal fistulas were associated with cirrhosis (Chapter VIII).

It has been suggested that in the long term the arterialization of portal flow can lead to periportal and perisinusoidal sclerotic lesions, which could in fact explain the portal hypertension (102, 103, 104). Some authors have noted that these lesions were primarily visible on electron microscopy (105) but these experimental results seem inconsistent.

The existence of portal hypertension by isolated increase of portal flow without any associated obstruction still remains controversial. If such observations exist, they seem to be quite rare and the association to an increase of portal flow of a discrete increase of the intrahepatic resistances sometimes secondary to the arterialization of the portal blood cannot be strictly eliminated.

However, the increase of the hepatopetal portal flow does aggravate a moderate portal hypertension and in fact reveal these minimal intrahepatic lesions. These lesions would have been too minimal to lead to a significant reduction of the portal flow and would have remained clinically latent or else revealed only quite late by a very large splenomegaly. This is probably the case of most of the portal arteriovenous fistulas as well as certain cases of portal hypertension with very large splenomegaly reported under the name of hyperkinetic portal hypertension.

7. CONTRIBUTION OF ARTERIOPORTOGRAPHY TO THE TREATMENT OF PORTAL HYPERTENSION

Surgical porto-caval anastomoses were for a long time the main therapeutic method of portal hypertension, allowing for a limitation in the risk of bleeding from esophageal varices.

Angiographic studies are, of course, necessary preoperatively to evaluate the possibility for the planned surgical shunt to be performed and postoperatively to check the shunt patency. Several angiographic methods may be tried but arteriography is particularly useful since it permits the evaluation of the morphological pattern of the shunt together with the resulting blood flow changes.

Three different problems will be discussed:
- The checking of surgical porto-caval anastomosis by arterioportography;
- The role of the preoperative hemodynamical disturbances in the choice of the therapeutic method;
- The possibilities of interventional radiology.

I. Checking of surgical porto-caval anastomosis by arterioportography

Direct opacification of the portal system or selective shunt catheterisation through the inferior vena cava may be more reliable methods than arterioportography to check the patency of a surgical porto-caval shunt. This is undoubtedly true from a morphological point of view since opacification of a porto-caval anastomosis either spontaneous or surgical, through arterioportography depends on its blood flow rate. Thus a patent surgical porto-caval anastomosis may not be visualized by arterioportography when narrowed enough for the blood flow rate through it to be markedly reduced. Therefore arteriography permits a better appreciation of the hemodynamical efficiency of the shunt, which is obviously the most important point, together with the induced changes in the hepatic and splenic blood flow (111).

Two different groups of surgical shunts may be performed: standard shunts, their main drawback being that they result in a markedly reduced hepatopetal flow; selective shunts, such as Warren's distal spleno-renal anastomosis, whose aim is to drain esophageal varices while preserving the portal inflow to the liver.

1. Standard shunts

After a standard porto-caval shunt (side-to-side or end-to side porto-caval anastomosis, proximal spleno-renal anastomosis or meso-caval shunt) the shunt patency is well demonstrated by evidencing opacification of the shunt together with inferior vena cava wheras the hepatofugal flow towards esophageal varices is no longer visualized (Figs. 64–69). Conversely, the lack of visualization of the shunt and inferior vena cava while opacification of the esophageal varices persists, proves that the shunt is not functioning either because it is occluded or markedly narrowed.

The shunt flow rate may be evaluated through arterioportography depending on its width and the degree of opacification of the inferior vena cava as compared to the portal vein opacification. The blood flow rate through the shunt is all the higher as the veins flowing toward it are more dilated and more densely opacified and the hepatopetal portal flow more reduced.

– *Following a side-to-side porto-caval anastomosis* interruption of the hepatopetal portal flow has been widely evidenced either by hepatic venography or arterioportography (112). It is associated with a widened hepatic artery and an atrophic liver. In fact, not only is the portal flow interrupted but it is reversed as the portal vein becomes a completely hepatofugal channel flowing towards the porto-caval shunt. Complete reversal of the intrahepatic portal flow is known to be a common finding following side-to-side porto-caval anastomosis.

Although the hepatic arterial flow rate is apparently all the more increased as the hepatofugal portal flow is higher, this reversed intrahepatic portal flow probably results in a decreased blood supply to the liver parenchyma, which might be responsible for the atrophy of the liver as well as the cases of post-operative hepatic failure which have been reported.

– *Proximal spleno-renal anastomosis* theoretically results in the same hemodynamical manifestations. However, due to its usually narrower diameter and its lower flow rate, it should be responsible for a lesser and more progressive reducing of the hepatopetal portal flow (Fig. 64).

In some cases, where successive arterioportographic checking may be performed, the portal inflow may be seen to decrease progressively, then to reverse a few months or years later whereas the increasing splenic vein diameter seems to accommodate to a progressive increase of the shunt flow rate (Fig. 65). Therefore, the proximal splenorenal anastomosis is perhaps less hazardous for the liver cell function since it probably allows for a better and more progressive adaptation of the liver blood flow to the new hemodynamical conditions.

– *Meso-caval interposition shunt* was proposed by Drapanas who anticipated that only the mesenteric venous blood would be diverted through the anastomosis whereas the splenic blood would continue to flow towards the liver (113, 114). It is, however, well demonstrated nowadays that such an anastomosis does not cause less hepatofugal portal outflow than a standard shunt (115, 116, 117).

Among 14 patent surgical mesocaval shunts checked by arterioportography the hepatopetal portal flow was found to be completely interrupted in 10 whereas the mesenteric and the splenic venous blood flowed through the shunt and then the inferior vena cava (Fig. 66). In one of these patients whose preoperative splenic blood flow rate was shown to be markedly slowed, the post-operative checking evidenced that the splenic circulation pattern returned to normal since the mesocaval shunt opened a new outflow tract for the splenic blood (Fig. 7).

In addition, 3 out of these 10 patients had a reversed intrahepatic portal flow demonstrated by retrograde opacification of the portal vein through a selective hepatic arteriography, which means that the arterial flow to the liver itself was partly diverted through the shunt together with the mesenteric and splenic venous blood (Fig. 67). In the four other cases, the portal vein was faintly opacified on the venous phase of splenic arteriography whereas the intrahepatic portal branches were not or nearly not visualized, which corresponds to a very poor or probably nearly interrupted portal flow. Therefore, the meso-caval interposition shunt seems to behave as a well-functioning side-to side portocaval shunt, permitting the mesenteric and splenic blood and sometimes the hepatofugal portal flow as well to drain towards the inferior vena cava.

According to some authors it could induce a lesser risk of hepatic failure than a truncular anastomosis. However, two-thirds of the patients operated on experienced more or less severe signs of encephalopathy (117).

Owing to its relatively high hepatofugal flow rate, the effectiveness of the surgical mesocaval shunt on preventing variceal hemorrhage is unquestionable. Therefore it could be postulated that a post-operative recurrent bleeding from esophageal varices results from occlusion of the shunt. Although this is probably true in most cases it is interesting to notice that a left-sided segmental portal hypertension has been encountered following meso-caval interposition shunt allowing for esophageal varices to be fed again from the splenic blood flow in spite of a perfectly patent shunt. In two patients who rebled shortly after a surgical meso-caval shunt, Witte et al. (118) by using angiography and pressure recording in the superior mesenteric vein and the splenic vein demonstrated effective decompression of the greater splanchnic venous system but continued gastro-splenic venous hypertension responsible for persistent esophageal varices. As a result of this, splenectomy permitted control of bleeding in both cases.

It is then demonstrated once more that contrary to what could be anticipated, hemodynamical changes resulting from an intrahepatic obstruction are not necessarily identical in the splenic vein as in the mesenterico-portal axis. On the contrary as already stated, the splenic venous flow rate is usually much more reduced than the superior mesenteric one when the total portal outflow is reduced.

In case of left-sided segmental portal hypertension, the mesocaval shunt flow rate is not high

enough to drain the whole portal outflow so that the mesenterico-portal blood only is diverted through the shunt. This results in a functional obstruction of the splenic vein responsible for a persistent splenic hypertension, forcing the splenic blood to flow away through short gastric veins and esophageal varices.

In three patients who had recurrent bleeding several months following a meso-caval interposition shunt, a left-sided segmental portal hypertension was clearly demonstrated by arterioportography (Fig. 68). Under these circumstances, the venous phase of superior mesenteric arteriography demonstrates the shunt to be normally patent whereas on coeliac arteriography the spleen pattern corresponds to a severe portal hypertension with a slowed splenic arterial flow. The splenomegaly remains moderate or minimal whereas its size usually progressively increases following surgery since the portal pressure is usually markedly lowered but not reduced to normal value by the porto-caval anastomosis. The arterial branches within the spleen are thin, the splenography is delayed and prolonged and the splenic vein is faintly visualized. The gastro-esophageal varices arising from the splenic hilum through short gastric veins are more or less faintly opacified since they usually have a low hepatofugal flow rate (Fig. 68) but this angiographic spleen pattern corresponding to a severe portal hypertension is quite unusual in a patient with a patent surgical meso-caval shunt and may be considered as typical for a left-sided segmental portal hypertension.

This gastro-splenic portal hypertension may exist immediately following surgery as in the cases reported by Witte. It is also likely to occur progressively as the result of the increasing intrahepatic resistances together with the hepatofugal portal flow rate. As a matter of fact, as soon as both superior mesenteric and portal hepatofugal flow rates are equal to the maximal shunt flow rate, a functional obstruction of the splenic vein occurs, giving rise to left-sided segmental hypertension.

Although probably very rare this segmental gastro-splenic hypertension deserves to be recognized so as to explain the possibility of recurrent bleeding despite a patent porto-caval shunt and the need for splenectomy as a therapeutic procedure.

2. Selective decompression shunts

The distal spleno-renal shunt described by Warren et al. (119) aims to decompress the esophago-gastro-splenic sector of the portal bed by permitting the varices to drain through short gastric veins and splenic vein towards the left renal vein while preserving the hepatopetal portal flow through the mesenterico-portal axis. Indeed the interruption of the portal flow to the liver is considered as the main cause of post-operative encephalopathy. As a matter of fact, the Warren anastomosis has proved to be as efficient as the standard anastomosis to prevent further bleeding from the esophageal varices without inducing such a high risk of liver insufficiency or neuropsychiatric disorders (119, 120). This is why this selective shunt is actually prefered by most authors.

The shunt patency is easily checked by the venous phase of splenic arteriography evidencing a good opacification of the left renal vein and inferior vena cava through the shunt since the splenic vein ends laterally into the left renal vein (Fig. 69). On the other hand, there is theoretically no modification of the mesenterico-portal axis as compared to the preoperative status.

Several works have shown that the distal spleno-renal shunt resulted in most cases in a progressively decreased hepatopetal portal flow (120, 121, 122, 123). According to Maillard, mean total hepatic blood flow was found at only 63% of preshunt level, 2 to 5 weeks after surgery, and collateral flow to the esophago-gastro-splenic sector had recurred in 17 of 18 patients (120). Other authors noticed that the portal vein flow was found to be hepatofugal in 6 of 16 patients checked by arterioportography (121).

It is then demonstrated that in most cases, new hepatofugal collateral pathways develop allowing for the portal blood to drain through gastric veins towards the spleno-renal anastomosis. These collateral veins uniting the left gastric vein to the left renal vein through short gastric veins are clearly documented on the venous phase of superior mesenteric arteriography several months or years following surgery (116, 122, 123).

Therefore, the Warren shunt appears to result in the long run in the same hemodynamical changes as the standard porto-caval anastomosis. However

the increased hepatofugal flow rate, as well as the decreased hepatopetal portal flow occur more progressively, accounting probably for a better adaptation of the liver blood supply. It is probably the reason why the incidence of encephalopathy remains low in all the series reported.

In fact, the Warren procedure aims to disconnect the high pressure mesenterico-portal axis, from a left-sided gastro-splenic axis where the pressure returns to normal following surgery. Because of the high pressure gradient between both venous systems, it is then normal for a collateral circulation to develop more or less quickly, even though the left gastric vein has been surgically ligated.

As a matter of fact, it was demonstrated by using hepatic venography that following an end-to-side porto-caval anastomosis, a hepatofugal collateral flow might occur from the hepatic end of the portal vein towards the superior mesenteric vein through the cystic and pancreatico-duodenal veins, permitting the intrahepatic portal blood to join the porto-caval anastomosis. (55, 56).

Since such hepatofugal collateral pathways may spontaneously develop following an end-to-side porto-caval anastomosis while the hepatic end of the portal vein is occluded, it is, of course, much more likely to occur when the portal vein is patent as is the case after a Warren procedure.

II. Treatment of portal hypertension according to hemodynamical changes

Two different groups of therapeutic methods may be used for the treatment or prevention of esophageal varices hemorrhage:

- lowering of portal hypertension by performing a surgical porto-caval shunt, or
- occluding venous collateral circulation feeding the varices by using endoscopic sclerosis, transhepatic embolization or various surgical procedures of devascularization.

Surgical porto-caval anastomoses have been widely used thus far and have proved to be efficient. The porto-caval shunt's efficacy to prevent recurrent bleeding is also well demonstrated in some cases of cirrhosis associated with a high flow rate spontaneous porto-caval anastomosis running through anatomical pathways other than gastro-esophageal veins.

The patients are prevented from bleeding at least for a long time and portal hypertension may remain inconspicuous despite a severe obstruction and a completely interrupted portal inflow. Owing to this, hepatic failure and encephalopathy appear to be the first signs of the disease at a late stage of atrophic cirrhosis. Unfortunately this result on variceal bleeding is obtained at the cost of diminished liver perfusion and an increased incidence of death due to liver insufficiency.

The vascularization of the cirrhotic liver parenchyma seems to depend greatly on the variations of the portal pressure and it is likely that a relatively high portal pressure is needed for a correct blood supply to the liver cells to be maintained. It may be then anticipated that any surgical procedure should induce as minimal changes as possible in the hemodynamics of the liver blood flow. Therefore, it is probably worth documenting as precisely as possible the preoperative hemodynamical abnormalities and particularly the portal flow rate and direction (124, 125, 126, 127).

1. Portal hypertension with moderately decreased portal inflow

In the cases where hepatic artery diameter is normal or near normal and the intrahepatic portal branches remain well opacified on the venous phase of S.M.A, an end-to-side porto-caval anastomosis which would result in immediate interruption of the portal flow to the liver is probably hazardous and should be avoided. Other standard porto-caval shunts are likely to have the same drawbacks although the portal inflow probably decreases more progressively following these surgical procedures. The Warren distal spleno-renal shunt seems then to be the most suitable procedure as generally agreed, since it preserves the hepatopetal portal flow, at least for a time.

2. Portal hypertension with markedly reduced portal inflow

The venous phase of S.M.A. shows a decreased opacification of the intrahepatic portal branches associated with a predominant hepatofugal flow

whereas hepatic artery diameter and arterial hepatography are greatly increased. Since the portal inflow is so much reduced that the liver parenchyma blood supply comes essentially from the hepatic artery one may wonder if the end-to-side porto-caval anastomosis is not a suitable procedure to be proposed.

Although the intrahepatic portal pressure may be somewhat reduced immediately following surgery, it is likely for the whole intrahepatic venous bed to be quickly filled from the arterial route through the arterio-portal shuntings, which are known to develop in cirrhotic liver. In certain cases, porto-splanchnic hepatofugal collateral pathways are seen to develop in the hepatic pedicle permitting some of the arterial blood supply of the liver to flow away through the porto-caval shunt (55, 56). It is then proved that owing to increased arterial flow rate, a high portal pressure persists within the liver following an end-to-side porto-caval anastomosis.

Such a hemodynamical situation closely likens the one resulting from a Warren shunt, when collateral pathways progressively develop allowing for the portal blood to drain towards the distal spleno-renal anastomosis through short gastric veins. However, the hepatofugal portal flow rate through this collateral network is probably lower following an end-to-side porto-caval shunt than after the Warren procedure since the portal vein is interrupted in the first case whereas it remains patent in the second.

For the lost portal inflow resulting from the end-to-side portocaval anastomosis to be compensated, an arterialization of the intrahepatic portal vein was proposed to be associated with the shunt procedure (128, 129). However, when an increased hepatic arterial flow compensates for a markedly reduced portal inflow as commonly seen in cirrhosis the usefulness of such a surgical arterialization of the portal vein appears to be questionable. In addition, by using arteriography to check the patency of this surgical arterio-portal shunt, Hammers reported the intrahepatic portal branches to have narrowed and a hepatofugal porto-splanchnic collateral circulation to have developed in most cases (128). Owing to these findings, it may be assumed that arterialization of the portal vein resulting in an improvement of the portal perfusion of the liver seems unlikely.

In the case of very poor hepatopetal portal flow the end-to-side portocaval anastomosis should perhaps be preferred to the other standard shunts through which a hepatofugal portal flow is more likely to occur because the portal vein patency is preserved. Under these circumstances a marked widening of the hepatic artery is seen to occur but one may wonder whether the arterial flow increases sufficiently for the hepatofugal portal flow through the surgical shunt to be compensated. It is then possible in some cases for a side-to-side porto-caval anastomosis to result in a 'steal syndrome' provided the intrahepatic arterioportal communications are developed enough to allow for a high hepatofugal portal flow rate.

3. Portal hypertension with hepatofugal portal flow

Once the intrahepatic portal flow is reversed, the portal vein turning out to be the main outflow tract of the liver, the risk for a surgical shunt to result in an impairment of the blood supply of the liver parenchyma is probably higher. As a matter of fact, either a side-to-side porto-caval anastomosis or a meso-caval or proximal spleno-renal shunt should result in an increased hepatofugal flow rate.

An end-to-side porto-caval anastomosis is probably less hazardous in case of low hepatofugal flow rate. However, when the hepatofugal flow rate is high, the surgical interruption of the portal vein may probably induce a quick rise of the intrahepatic portal pressure resulting in a decreased hepatic arterial flow and sometimes in hepatic ischemia. In fact, most of the cases associated with a completely hepatofugal portal flow correspond to a late stage of the disease associated with a pronounced liver insufficiency as evaluated by Child's classification. For this reason many of these patients are considered poor candidates for shunt surgery.

Since the liver blood flow disturbances essentially depend on the anatomical lesions involving the liver it may be thought that in cirrhosis the more damaged the liver parenchyma, the more severe the hepatic failure and the more reduced the hepatopetal portal flow. This is probably why most patients considered as good candidates for surgery according to Child's classification belong to the group where the Warren procedure seems to be the

most suitable since they have a persistent hepatopetal portal flow.

The relationship between the portal flow direction and the Child's classification was studied in a group of 234 cirrhotic patients treated by percutaneous transhepatic embolization of esophageal varices. A statistically significant correlation ($X^2 = 22.7$) was found to exist between either the persistent hepatopetal portal flow and Child's class A and B, or the reversed flow in the portal vein and Child's class C13, 14, 15. Conversely, there was no statistically significant correlation between the portal flow direction and Child's class C10, 11, 12 (130).

It is then demonstrated that in cirrhosis a completely hepatofugal portal flow signifies a severe hepatic failure.

In any case the chances of survival of a patient with a portocaval shunt depends not only on the hemodynamical changes but on the degree of hepatic insufficiency as well.

According to some authors, angiographic evaluation of the portal flow rate and direction has not proved as valuable as clinical signs and liver function tests in predicting the optimal shunt procedure and the chance of complications following surgery (98, 126). As a matter of fact, important circulatory changes such as either a completely interrupted portal flow or a reversed one may sometimes be well tolerated by the liver provided the pathological process results in little involvement of the liver cells.

In two patients with idiopathic portal fibrosis an interrupted or reversed portal flow was seen to be associated with few liver function tests abnormalities and a long survival (Fig. 44). In cirrhosis, however, it always seems possible for a quick change in the hemodynamics of the liver blood flow to result in a worsened liver cell insufficiency. The results of shunt surgery are therefore often considered as discouraging and devascularization procedures are gaining ground (131, 132, 133).

Various surgical procedures aiming to suppress the blood flow through the varices have been advocated – esophageal transection, gastro-esophageal resection, Sugira procedure, resection-anastomosis of the supracardial esophagus associated with left gastric vein and splenic artery ligation. By using this procedure a 55% five-year survival was reported, post-operative deaths excluded (133, 134). Finally, splenectomy might be advocated when a high splenic blood flow rate is supposed to play a role in portal hypertension.

It might be the case when a pronounced splenomegaly associated with either a post-hepatitis cirrhosis or a portal fibrosis raises the discussion about a 'hyperkinetic' portal hypertension. In fact, gastro-esophageal varices are sometimes seen to opacify only on the venous phase of coeliac arteriography whereas they are not visualized through the venous phase of S.M.A. which suggests they are fed mainly from the splenic venous outflow (Figs 70 & 71).

The left-sided segmental portal hypertension following a meso-caval interposition shunt is another condition when the need for splenectomy is advocated. However splenectomy is rarely resorted to in portal hypertension. In addition, according to some authors it could induce a risk of portal vein thrombosis (127).

III. Interventional radiology and portal hypertension

Owing to the liver failure, many cirrhotic patients are poor candidates for surgery. It is particularly true in actively bleeding patients and Child's class C patients in whom surgical treatment is often considered as contra-indicated because of a high post-operative mortality rate. For this reason, various non-surgical therapeutic procedures have been developed, the aim of which being either to suppress the flow through esophageal varices or to reduce the splenic blood flow or even to lower the portal pressure.

1. Transhepatic obliteration of esophageal varices

Described by Lunderquist in 1974 (35), this method seemed at first promising, then appeared to result only in a temporary occlusion of the varices in most cases. In addition, it was reported as technically difficult to perform and carrying a significant complication rate so that dissatisfaction with this procedure seems now widespread.

According to a recent report, transhepatic embolization of esophageal varices permits neither

the immediate or long-term mortality rate nor the recurrent bleeding rate to be reduced (136). In fact, results relying on too few cases are likely to explain such a pessimistic opinion since the results of this procedure depend greatly on the degree of the hepatic failure as is the case for every therapeutic method in cirrhosis. Other authors reported this procedure to be relatively efficient in the management of esophageal varices bleeding (137, 138).

After performing 370 transhepatic obliteration of esophageal varices in 326 patients for five years this method proved to be useful as a palliative treatment of variceal bleeding in cirrhotic patients (140). Due to increasing experience, technical failure rate and severe complications decreased by about 13% in 1980 to 2.5% in 1983.

Among 169 actively bleeding patients, the bleeding stopped in 85% of the cases (93.5% in Child's class B and 80% in Child's class C patients) so that immediate result on the bleeding is not questionable. However, some patients die within the first ten days from recurrent bleeding or liver failure so that 80% of the patients only survive at that time, the mortality rate depending essentially on the degree of hepatic failure (6% in Child's class B, 28% in Child's class C10–11–12, 48% in Child's class C13–14–15) (Figs. 32, 70–71).

Results on recurrent bleeding may appear discouraging as 56% of the patients rebleed within the first 6 months following embolization, with great variations according to the stage of the cirrhosis: 46.5% in Child's class B, 63% in Child's class C10–11–12, 86% in Child's class C13–14–15 patients). However, in about half the cases, recurrent bleeding is benign, responding well to medical care, so that 68% of the patients are alive at 6 months and 52.5% after one year (88% and 72% in Child's class B, 53% and 40% in Child's class C10–11–12, 50% and 32% in Child's class C13–14–15).

Transhepatic embolisation of esophageal varices may therefore be a relatively simple and safe procedure provided its technical difficulties are overcome by increasing experience. Its immediate efficiency on bleeding is unquestionable. The recurrent bleeding rate depends greatly on the stage of the disease and the degree of hepatic failure (140).

Unlike many other therapeutic procedures, transhepatic embolization of esophageal varices can be performed in every actively bleeding patient. It permits the amount of blood transfusion to be reduced and the worsening of hepatic insufficiency commonly resulting from bleeding to be checked. In patients considered good candidates for surgery, interruption of the hemorrhage allows for the patient to be operated on under better conditions. In inoperable cases embolization appears as an emergency procedure allowing for immediate and sometimes long-term survival, although rebleeding from recurrent varices does occur within a variable and often unpredictable period of time. For these reasons, transhepatic embolization remains according to some authors and in our opinion, a valuable tool for the treatment of esophageal varices bleeding.

2. Splenic artery embolization

Splenic artery embolization which results in nonsurgical splenectomy may be theoretically useful either to alleviate symptoms of hypersplenism or to control bleeding from esophageal varices by reducing the portal flow rate. According to some authors, splenic flow does contribute approximately 20 to 40% of portal flow. Removal of this inflow might reduce portal pressure (141, 142). In fact, surgical splenectomy as well as ligation or embolization of the splenic artery have been reported to control bleeding in certain patients with cirrhosis (143, 144, 145, 149).

However, in early reported cases splenic artery embolization appeared to induce a high risk of splenic abscess (150). The technique of partial splenic embolization was therefore advocated. Using sequential splenic embolization some authors reported minor complications in comparison to those undergoing complete embolization of the spleen (146, 147).

An occlusion similar to a surgical ligation was reported to be obtained by using Bucrylat, owing to its rapid polymerization and solidification (148). However, the same result seems easier to achieve by placing a stainless steel coil in the splenic artery. As a result of this, peripheral arterial branches in the spleen remain patent and function as channels for collateral circulation from gastric arteries. This collateral flow may be sufficient to reduce the possibility of an ischemic condition great enough to

precipitate anaerobic bacterial abscess formation.

Splenic artery embolization allowed Goldmann et al. to control variceal bleeding immediately in four cases of splenic vein thrombosis and eight cases with generalized portal hypertension. However, wedged hepatic pressure obtained in 12 patients immediately before and after arterial occlusion showed no changes (148).

In four out of seven cirrhotic patients who underwent splenic artery embolization with steel coils because of recurrent bleeding following transhepatic obliteration of esophageal varices, this procedure proved to be useful (Fig. 70). In some cases, however, it was impossible to obtain a complete occlusion of the splenic artery by using embolization with stainless steel coils (Fig. 71).

Three patients survived 18 to 21 months following embolization without recurrent bleeding. In the fourth patient frequency and severity of variceal hemorrhage was greatly reduced with an 18 month follow up.

The procedure was not beneficial in three other patients. One of them died from hepatic failure and in the two others endoscopic sclerosis or surgery were performed because of recurrent bleeding.

Splenic ischemia resulting from splenic artery embolization varies, depending on the spleen size as well as the development of the collateral circulation. It is likely that splenic ischemia will be all the more important as the spleen becomes more enlarged, as the collateral flow is relatively less efficient.

Under these circumstances, embolization may induce a prolonged left flank pain and a transient renal insufficiency, but no infectious complication was encountered (Fig. 70). Therefore, splenic artery embolization may probably be useful in the treatment of esophageal varices bleeding but only in the cases where varices development proves to result mainly from the splenic outflow. Indeed, this is a relatively rare hemodynamical condition as in portal hypertension, the splenomegaly results from portal blood stasis and the splenic blood flow rate all the less decreased as the portal hypertension is higher.

Splenic artery embolization may be advocated when arterioportography demonstrates the splenic venous outflow feeding the varices, which corresponds to four main circumstances:

1. Segmental splenic venous hypertension resulting from either splenic vein thrombosis or thrombosis of a surgical distal spleno-renal shunt (148).
2. Left-sided segmental portal hypertension following a meso-caval interposition shunt (118).
3. Hyperkinetic portal hypertension which usually corresponds to a minimal and long-standing intrahepatic obstruction allowing for a pronounced enlargement of the spleen. It is then possible for an increased splenic venous outflow to determine a slight increase of the portal pressure. In fact, arterioportography may prove the mesentericoportal flow to be essentially hepatopetal on the venous phase of S.M.A. whereas varices are opacified only through the venous phase of splenic arteriography (Figs. 68, 70, 71).

   From these arteriographic findings it may be concluded that owing to the reduced intrahepatic venous bed, only the mesenteric blood is able to flow away through the liver while the splenic venous blood does find collateral hepatofugal ways through esophageal varices.

   This hemodynamical condition is exactly the same as the left-sided segmental portal hypertension described after a meso-caval interposition shunt.
4. Recurrent bleeding following transhepatic obliteration of esophageal varices when resulting from development of new varices through short gastric veins. Since these veins join the splenic vein close to the splenic hilum it is likely for the splenic venous outflow to contribute to feed these varices. Splenic artery embolization may then be performed as a complementary procedure to the transhepatic embolization of the varices as was done in seven patients (Figs. 70 & 71).

Finally, the cases when splenic blood flows through esophageal varices at such a rate as to influence variceal bleeding seem relatively rare. However, when such a hemodynamical condition is suggested through arterioportography, splenic artery embolization may be performed, as it seems to be efficient in some cases, although there has been little experience with this method so far.

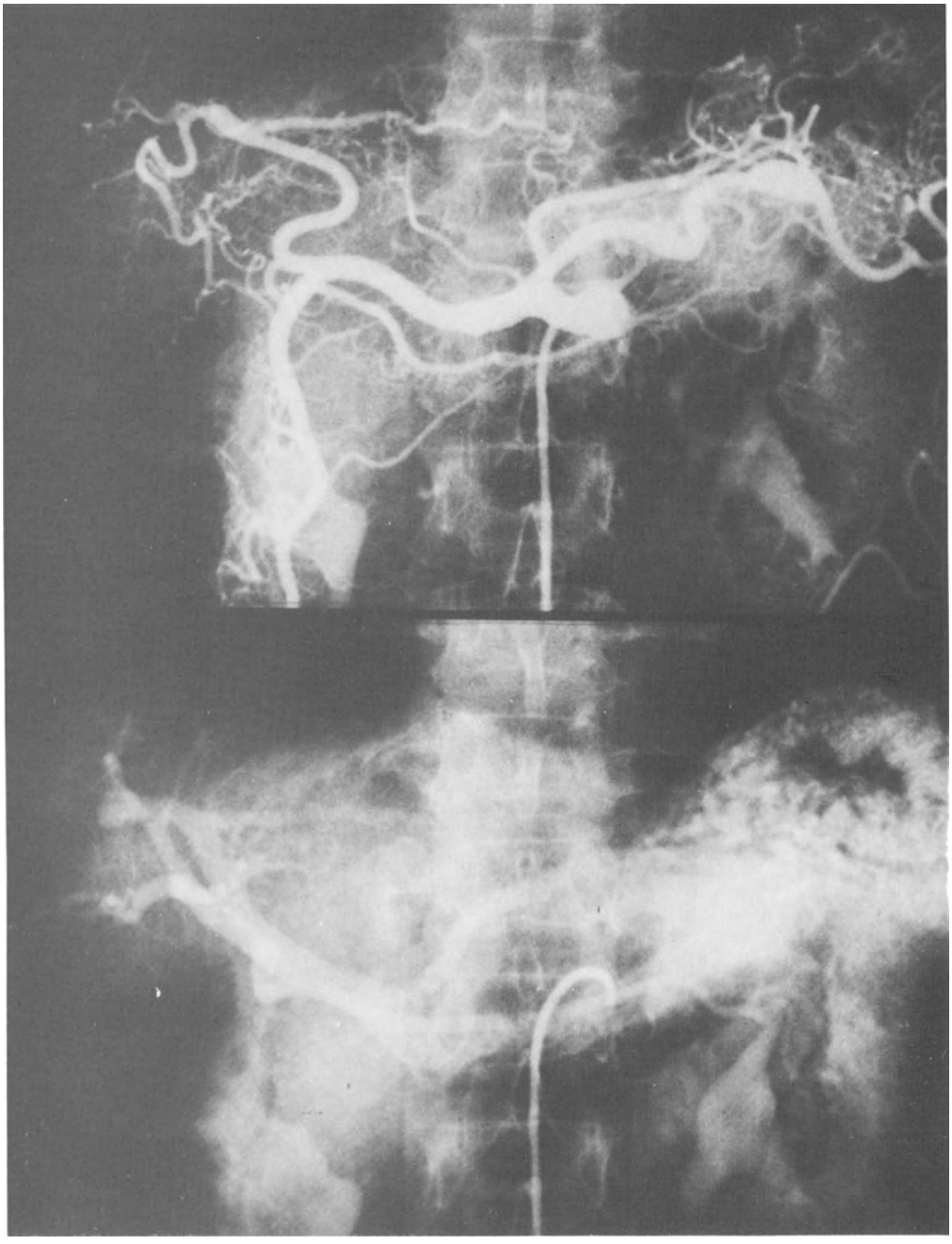
3. Other non-surgical therapeutic methods

Two other methods can be used for the management of variceal bleeding 1) Vaso-pressin infusion in the superior mesenteric artery, which by decreasing the portal flow control the bleeding in 70% of the cases according to some authors (152); and 2) Endoscopic sclerosis of esophageal varices (153, 154).

More recently, formation of intrahepatic portosystemic shunts using a balloon dilatation catheter was reported as a feasible procedure (155), but this method probably carries a significant venous thrombosis rate and its long-term efficacy is not yet proved.

Finally, many surgical or non-surgical therapeutic methods were advocated to control hemorrhage in portal hypertension. The choice of the most suitable one is contigent on three main factors: the primary disease, the degree of hepatic failure and the changes in the hemodynamics of the liver blood flow.

Non-surgical procedures are useful either as a palliative procedure in patients considered to be poor surgical candidates or as a preoperative method in actively bleeding patients. It is sometimes beneficial for these various procedures to be associated as they act through different mechanisms.



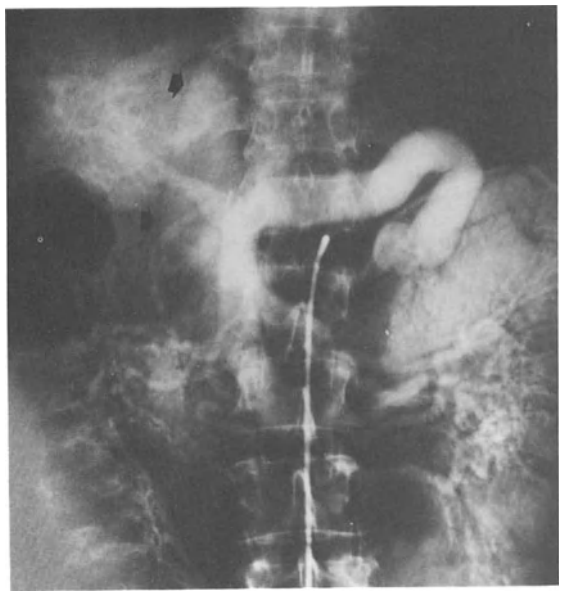
64a

Fig. 64. Angiographic checking of a proximal spleno-renal anastomosis performed 5 years earlier (1973) for post-hepatitis cirrhosis in a 63-year-old female.

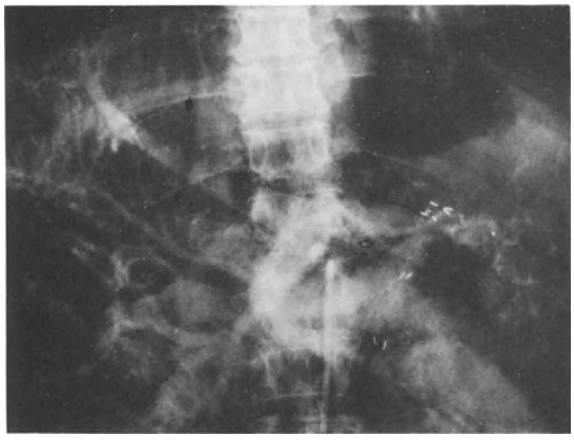
64a: Coeliac arteriography (16–5–78).

Atrophic liver with dilated hepatic artery and retrograde opacification of the portal vein seen 5 sec later which signifies the reversed intrahepatic portal flow.

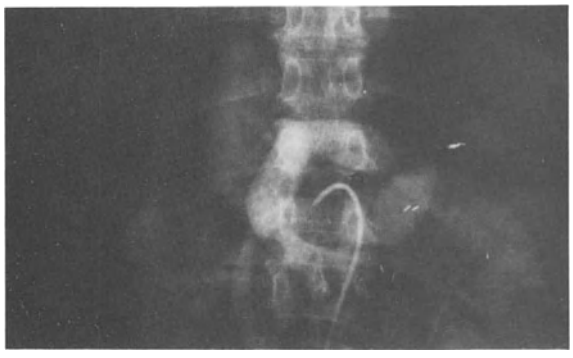
64b



64b: Venous phase of S.M.A.
Hepatofugal flow through the markedly widened splenic vein and the spleno-renal shunt towards inferior vena cava (↑).
– Portal vein nearly not visualized.



65a

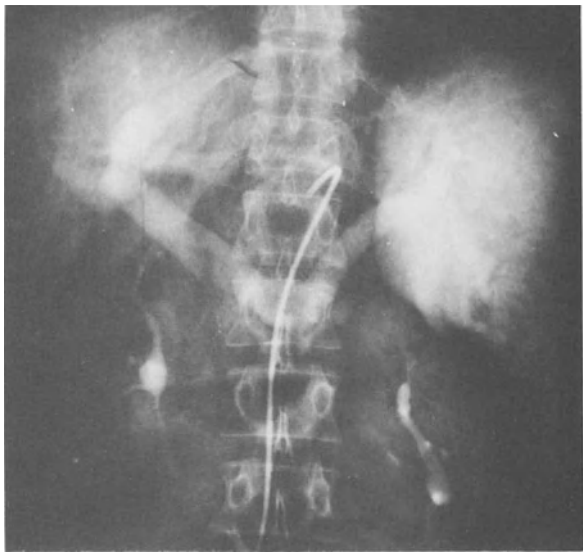


65b

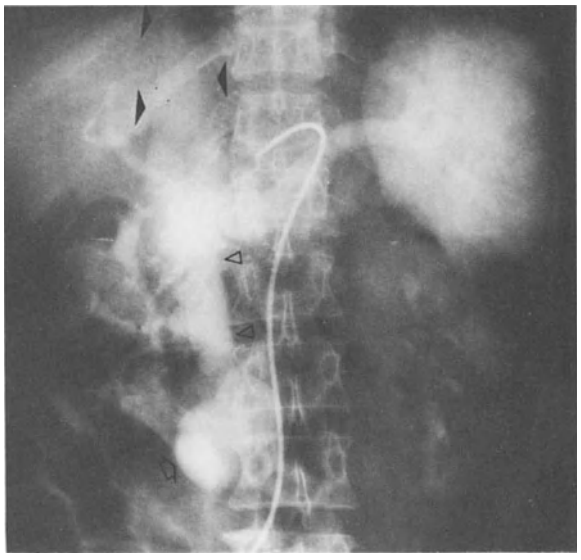
Fig. 65. Proximal spleno-renal anastomosis performed in January 1973 in a 58-year-old female with atrophic alcoholic cirrhosis.

65a: Angiographic checking 5 months following surgery (15–6–73).
Opacification of the inferior vena cave accounting for patency of the surgical shunt (↑).
The portal vein remains visible but hepatoportal flow is probably very poor.

65b: Angiographic checking 5 years later (2–12–78):
- The portal vein is no longer visualized (hepatic arteriography showed the intrahepatic portal flow to be reversed).
- Completely hepatofugal portal flow through the spleno-renal shunt with widening of the splenic vein as compared to the previous checking (⌓).
- The patient is still healthy 11 years following surgery.



66a



66b

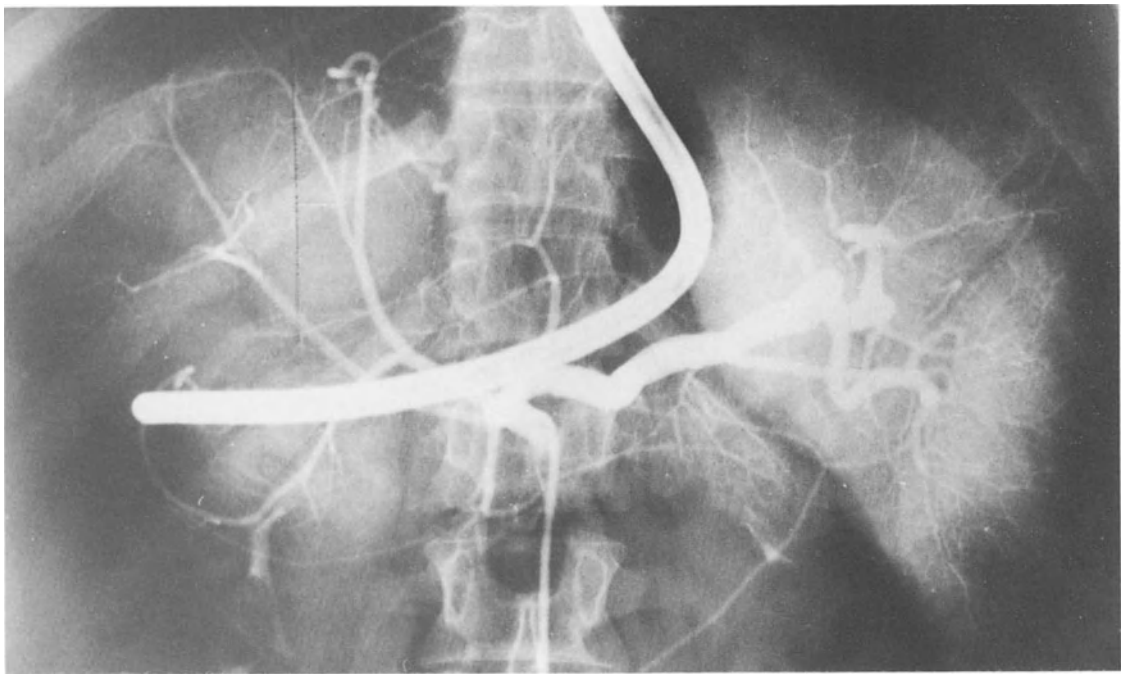
Fig. 66. Meso-caval interposition shunt performed in May 1976 in a 57-year-old female known to have alcoholic cirrhosis for 5 years. Corrected sinusoïdal pressure: 40 cm saline.

66a: Preoperative coeliac arteriography (16–4–76) shows a good blood flow through a moderate splenomegaly (16 cm) as the splenic and portal veins are well opacified, accounting for a probably moderate obstruction.

66b: Post-operative angiographic checking $1^1/_2$ years later (8–11–77) (venous phase of coeliac arteriography). Hepatopetal flow is no longer present.

The splenic vein drains retrogradely through the superior mesenteric vein (△) to join the meso-caval shunt (⌓) and inferior vena cava (▲).

Owing to the probably high splenic outflow rate through the shunt, the splenomegaly decreased (13 cm).

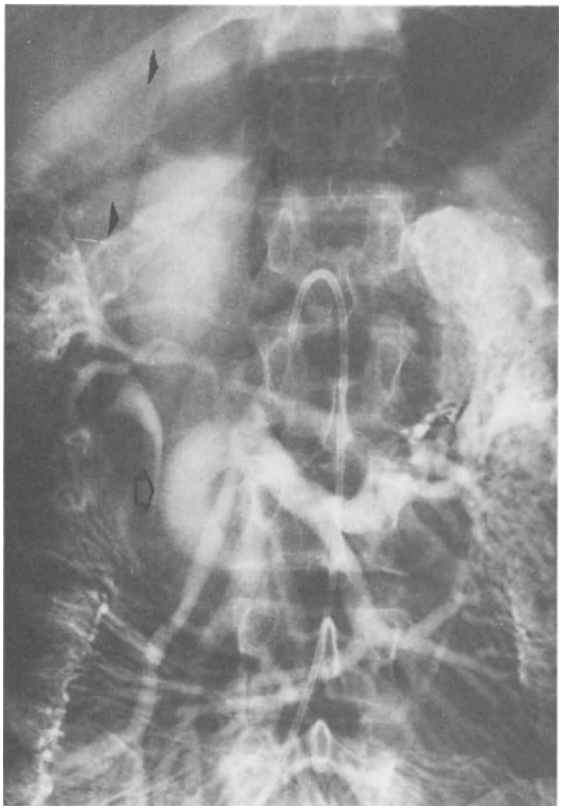


67a

Fig. 67. Meso-caval interposition shunt performed in December 1976 in a 26-year-old male with post-hepatitis cirrhosis.

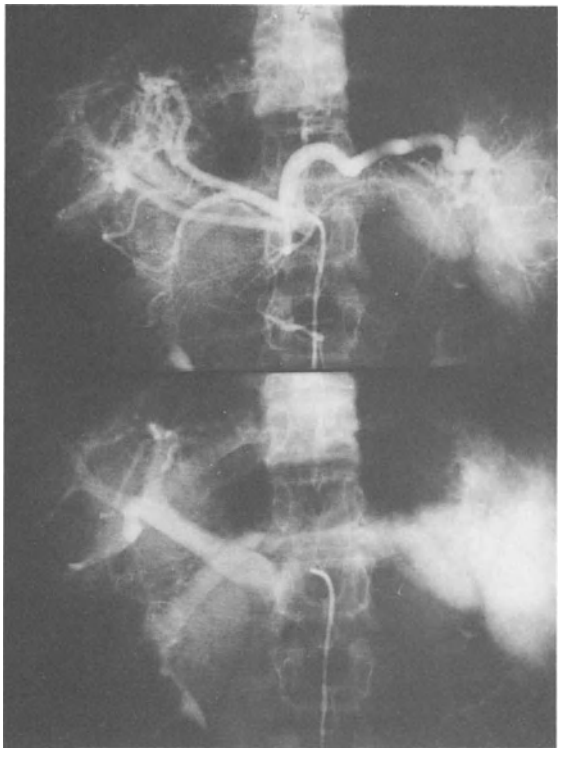
67a: Preoperative coeliac arteriography (1–12–76). Pronounced splenomegaly (20 cm) associated with atrophic liver and very thin intrahepatic arterial branches, i.e. the common angiographic pattern of post-necrotic cirrhosis.

67b: Post-operative angiographic checking 3 months later (16–3–77).
Patency of the shunt (⌓) is documented by the venous phase of superior mesenteric arteriography with a dense opacification of the inferior vena cava (▲).

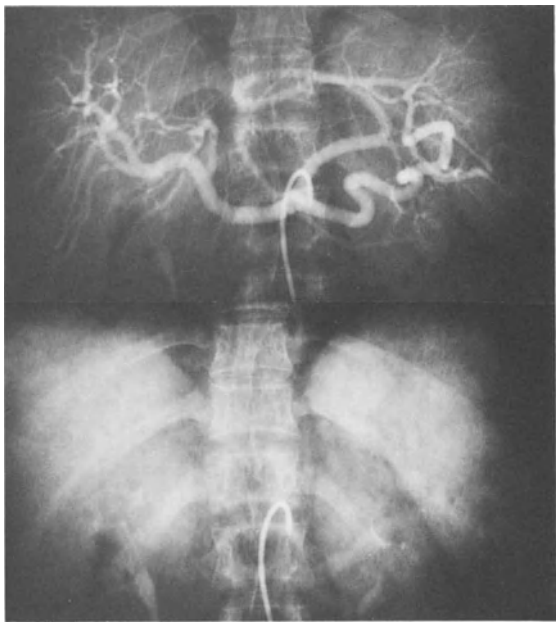


67b

67c



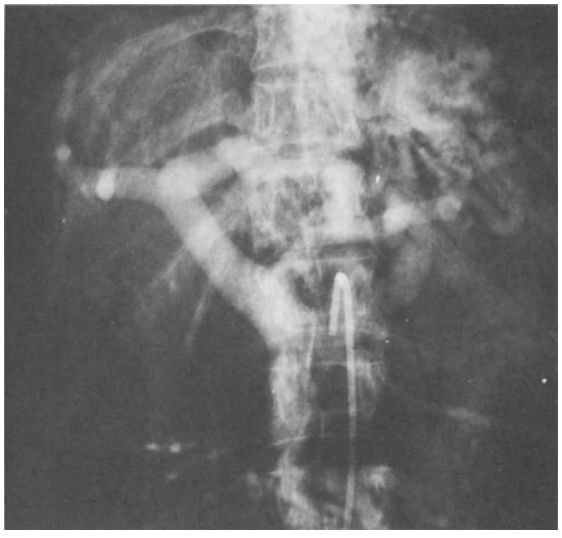
67c: Coeliac arteriography shows a marked dilatation of the hepatic artery to have occurred together with a massive arterio-portal shunting seen at the 4th sec.
Complete reversal of the intrahepatic portal flow. Moderate decrease of the spleen size (17 cm).
The patient is doing well 4 years following surgery.



68a

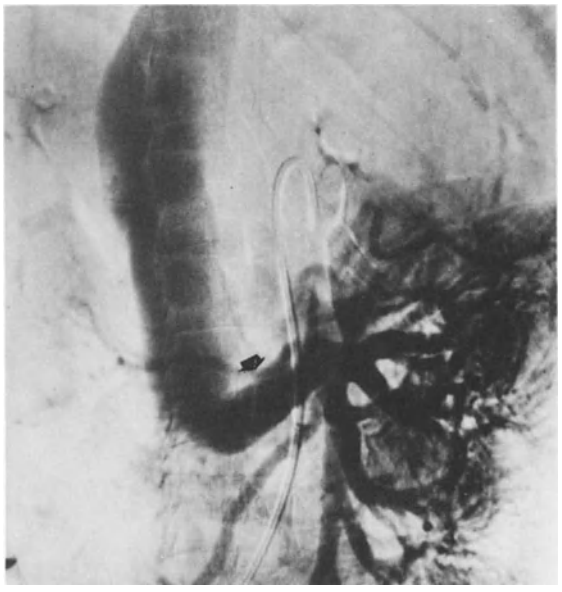
Fig. 68. Left-sided segmental portal hypertension after a meso-caval interposition shunt performed in January 1976 for alcoholic cirrhosis in a 30-year-old male.

68a: Preoperative arteriography (12–9–75).
Minimal splenomegaly (15 cm) with thin intrasplenic arterial branches and faint opacification of the splenic vein associated with hepatomegaly and markedly dilated hepatic artery.
Angiographic pattern of a severe and rapidly progressive intrahepatic obstruction.



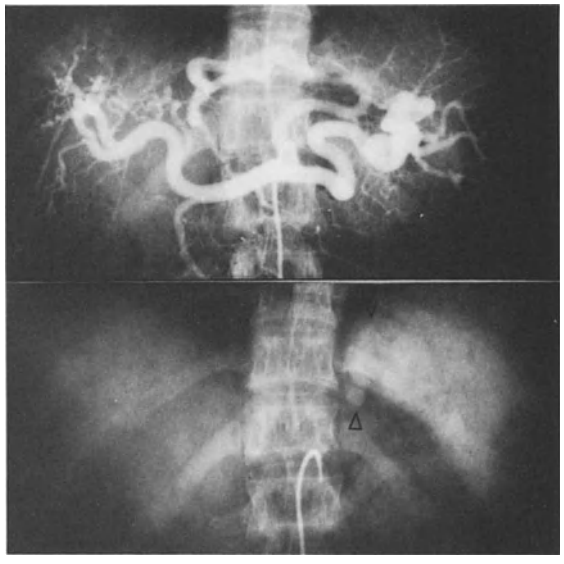
68b

68b: Venous phase of S.M.A. shows a poor hepatopetal portal flow and massive retrograde filling of gastro-esophageal varices through a wide left gastric vein (12–9–75).



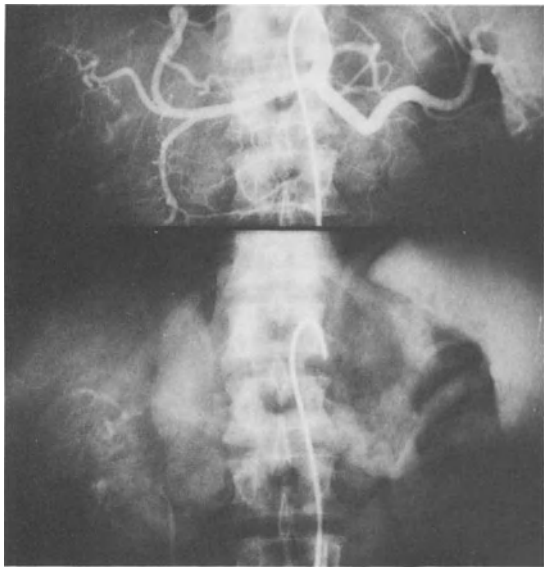
68c

68c: Angiographic checking for massive recurrent bleeding from esophageal varices 4 years later (January 1979).
Venous phase of S.M.A. (26–1–79) demonstrates the meso-caval shunt to be patent with dense opacification of the graft (↑) as well as the inferior vena cava.



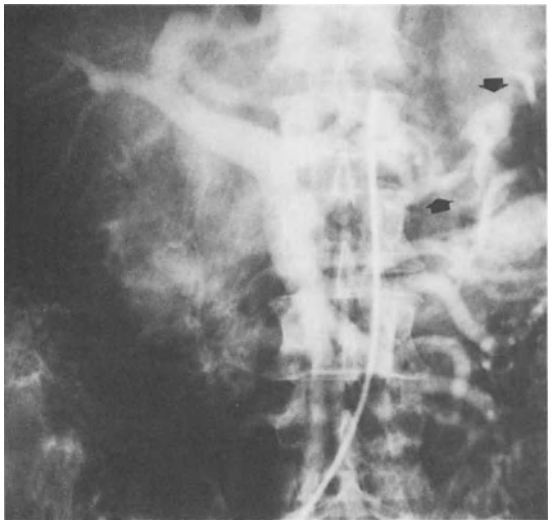
68d

68d: Coeliac arteriography (26–1–79) shows marked atrophy of the liver and increased widening of the hepatic artery.
The low flow rate pattern of the spleen persists without any change as compared to the preoperative period (Fig. 68a) (spleen size: 15 cm and faint opacification of the splenic vein). However, gastric varices are shown to opacify, probably from short gastric-veins (△).



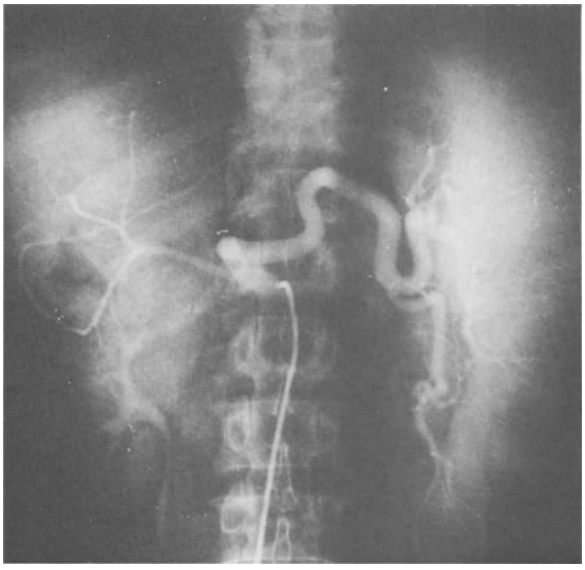
69a

Fig. 69. Warren's distal spleno-renal shunt performed in April 1978 in a 52-year-old cirrhotic patient.
Angiographic checking 8 months later (20–12–78).
69a: Coeliac arteriography
- only minimal dilatation of the hepatic artery
- near normal-sized spleen (14.5 cm) while the pre-operative size was 16 cm
- the splenic vein drains towards left renal vein and inferior vena cava demonstrating the shunt to be normally patent.



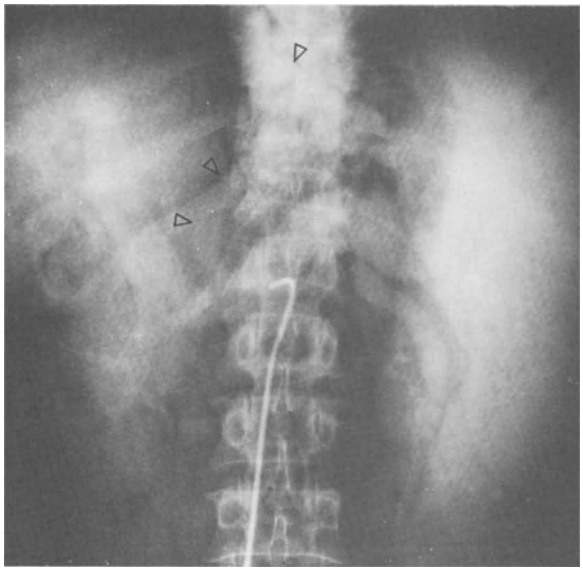
69b

69b: Venous phase of S.M.A.: relatively good opacification of the narrowed intrahepatic portal branches which signifies the moderately reduced portal inflow.
However, new hepatofugal collateral pathways are seen to develop (↑).



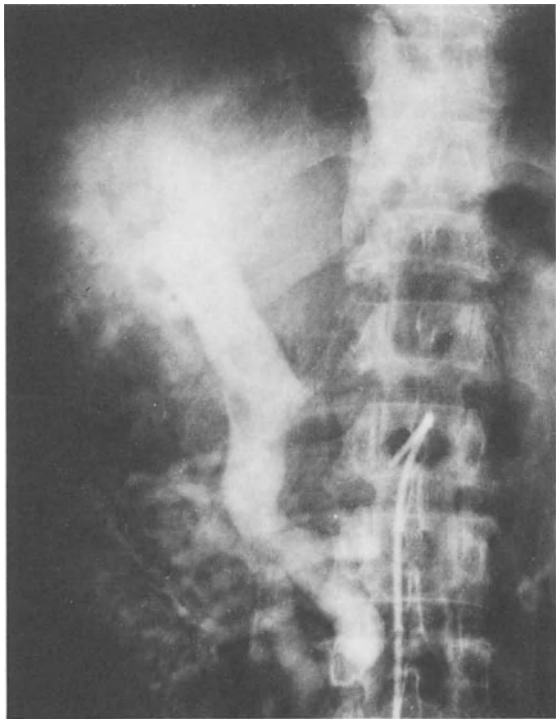
70a

Fig. 70. Post-hepatitis cirrhosis in a 56-year-old male from North Africa. Patient referred for splenomegaly, hypersplenism and large gastric varices (16–10–82).
70a: Huge splenomegaly with dilated and lengthened splenic artery associated with atrophy of the liver and very thin intrahepatic arterial branches. Angiographic pattern suggesting a long standing and slowly progressive portal hypertension due to either post-hepatitis cirrhosis or periportal fibrosis.

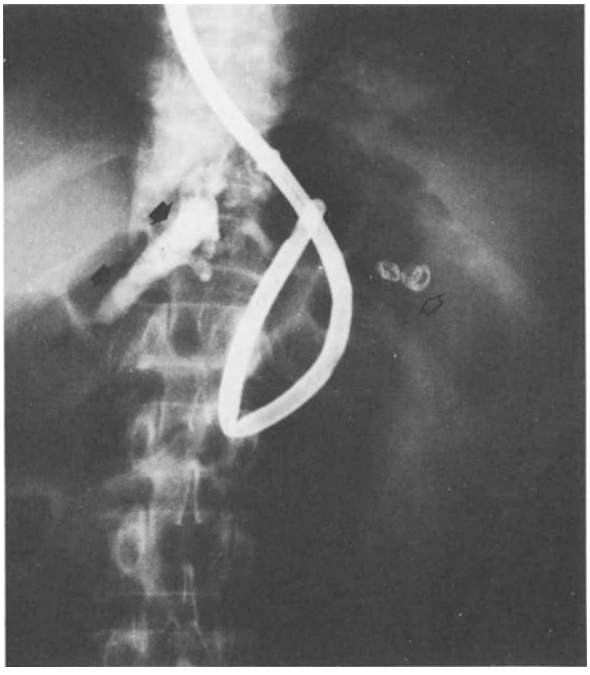


70b

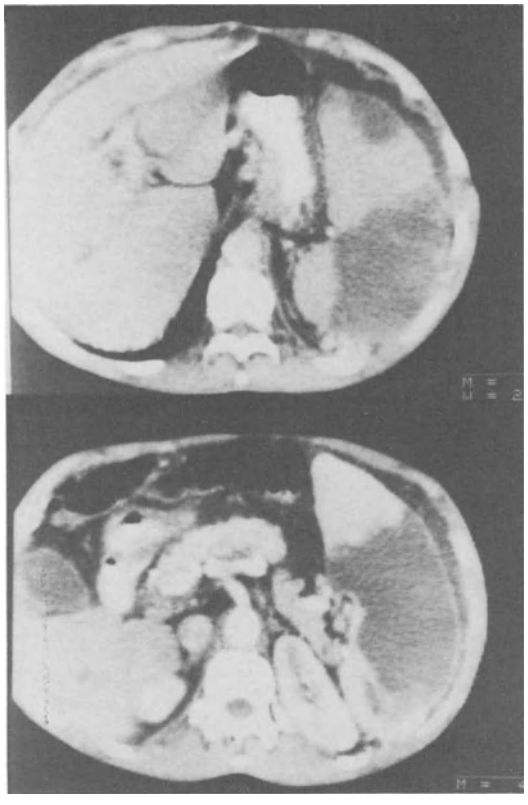
70b: Faint opacification of the markedly widened splenic vein and retrograde flow through the left gastric vein and varices faintly visualized (△).



70c



70e

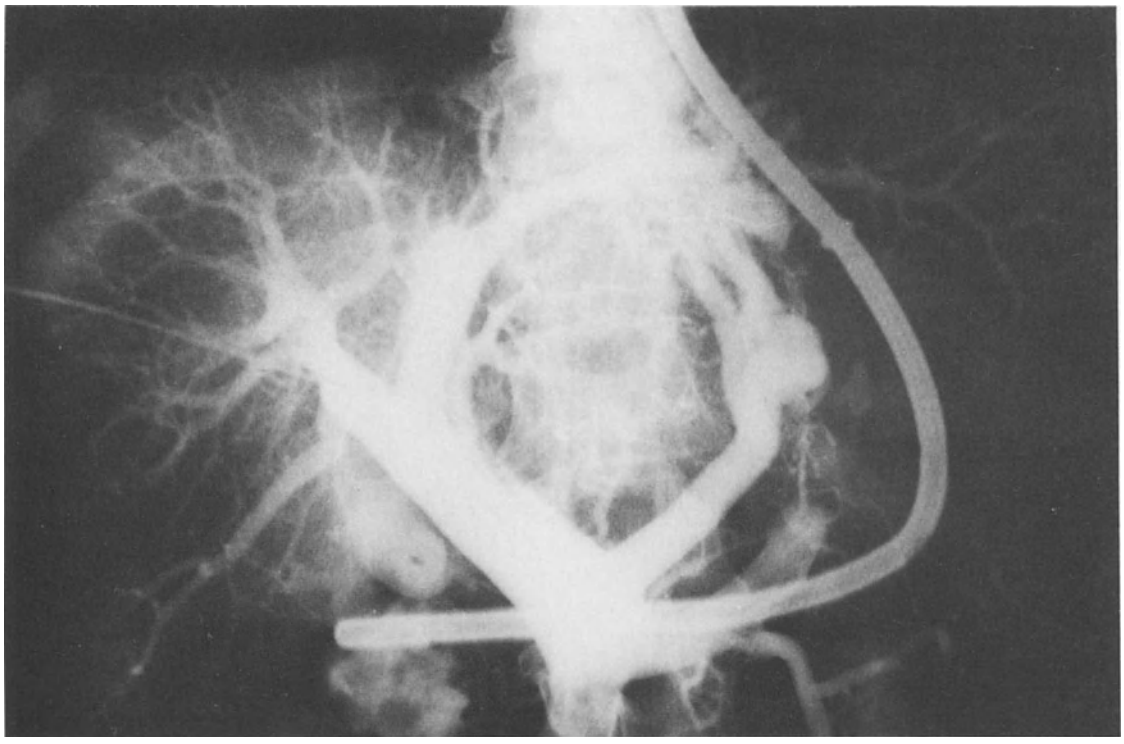


70d

70c: Venous phase of S.M.A.: hepatopetal flow through the dilated portal vein whereas no hepatofugal flow is seen. The left gastric vein is not visualized whereas it was opacified on the venous phase of splenic arteriography.
For this reason and because of hypersplenism, splenic artery embolization with three steel coils was performed (15–11–82).

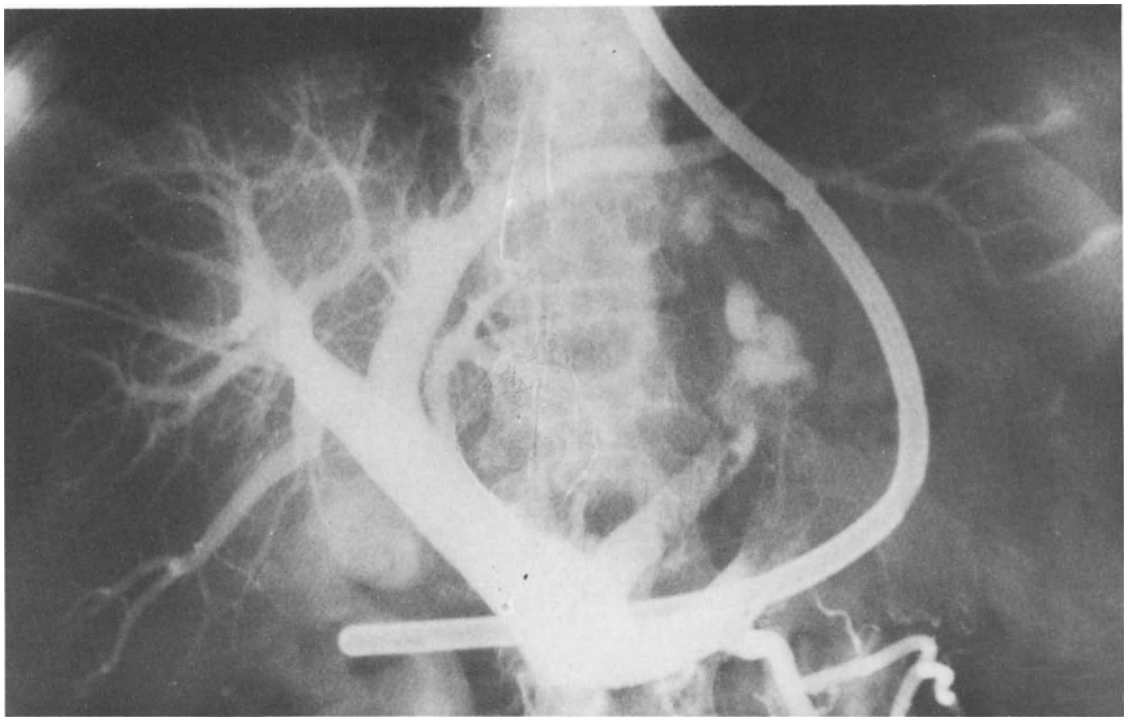
70d: Angio C.T. Scan (13–12–82) demonstrates partial infarctus of the spleen following embolization.

70e: Because of recurrence of bleeding four days later, percutaneous transhepatic obliteration of esophageal varices is performed (20–12–82).
Plain film of the abdomen following varices embolization shows the bucrylat mixed with lipiodol occluding the left gastric vein (◓) as well as the steel coils in the splenic artery (⌓).
Bleeding and hypersplenism remain controlled one year later (12–1–84).



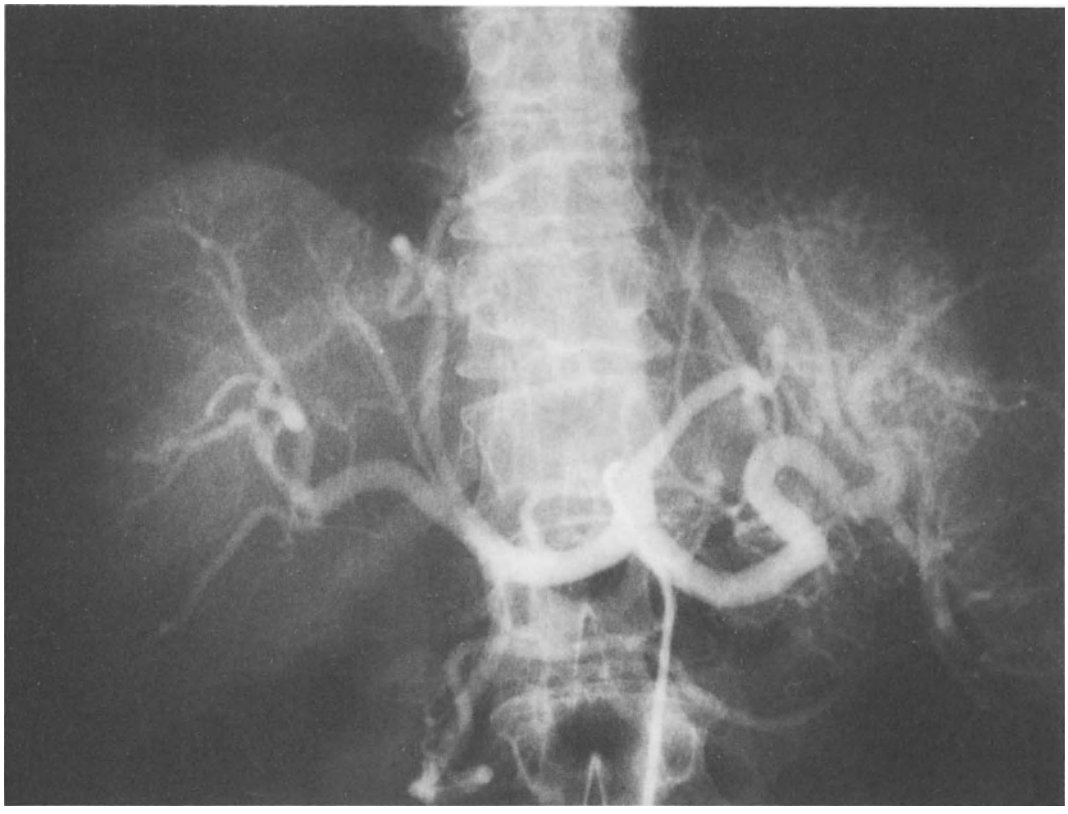
71a

Fig. 71. Alcoholic cirrhosis in a 48 year-old male treated by esophageal varices and splenic artery embolization.
71a: Percutaneous transhepatic portography performed as an emergency procedure (15–4–82) (actively bleeding patient belonging to Child's class C13).
Hepatofugal flow through a wide left gastric vein and esophageal varices.



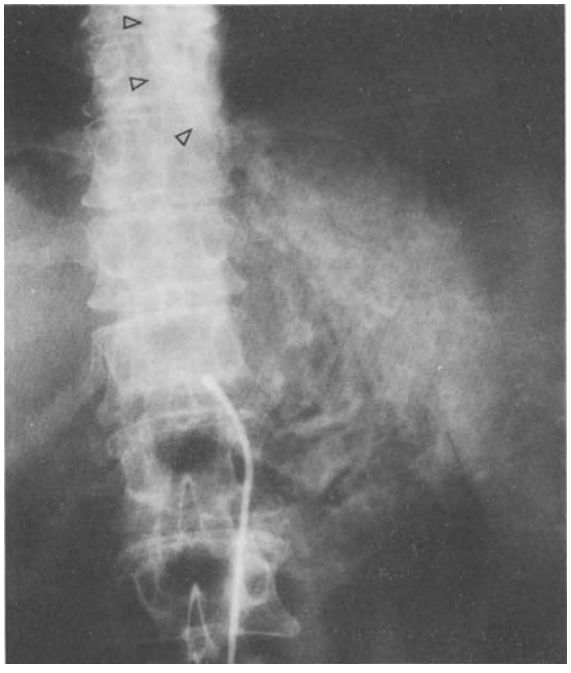
71b

71b: Following embolization with bucrylat mixed with lipiodol the left gastric vein is occluded.
Patient's condition and Child's score improved (B9) but a moderate variceal bleeding recurred 5 months later.



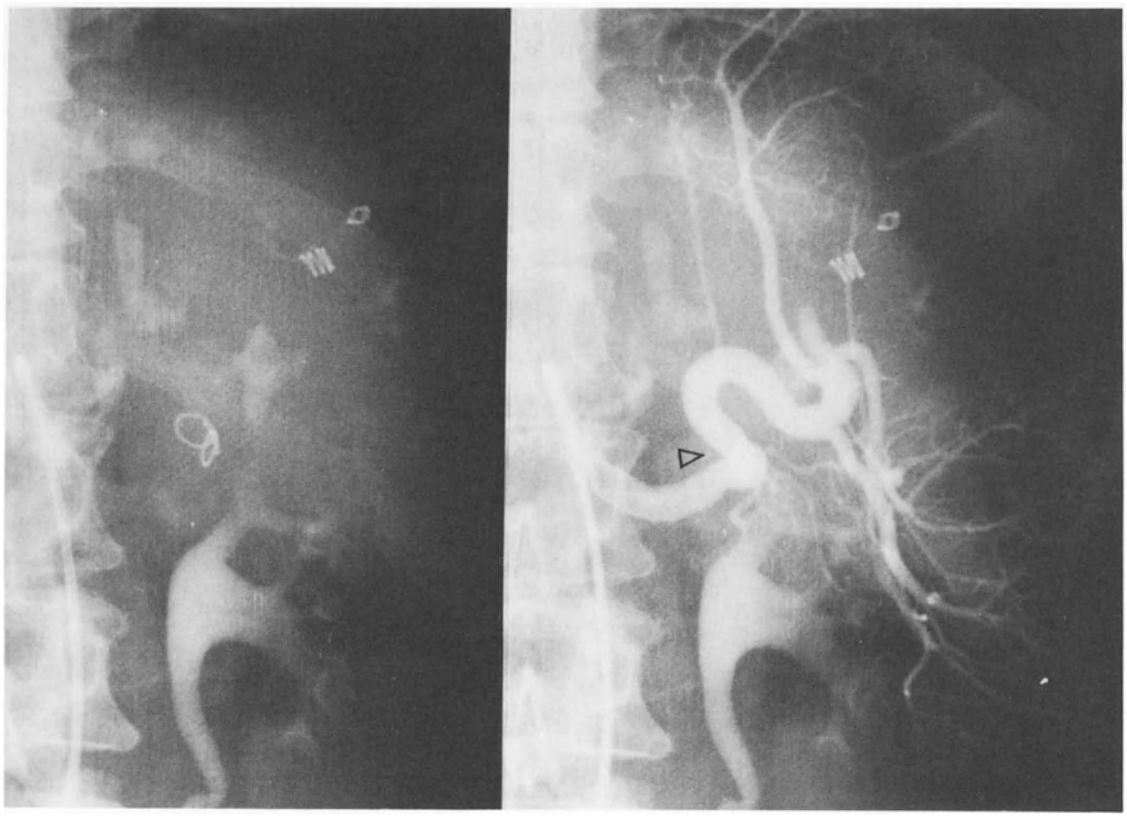
71c

71c: Arteriography (14–9–82) shows a dilated hepatic artery and a moderate splenomegaly (19 cm).



71d

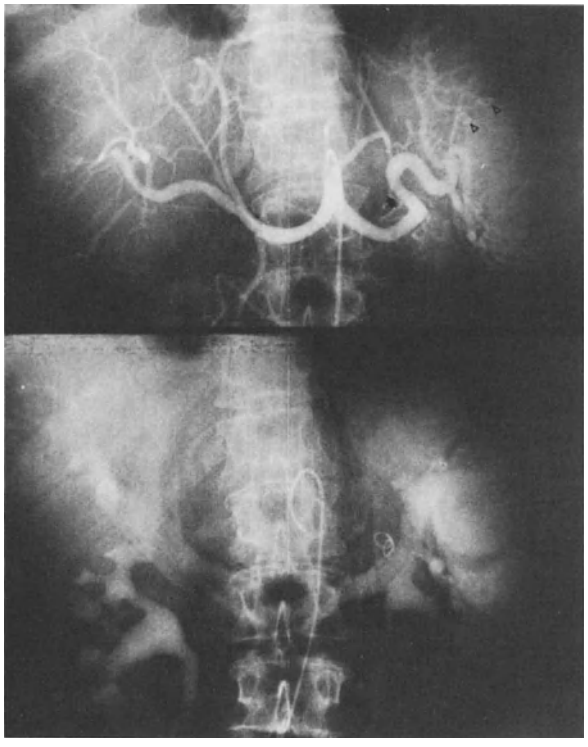
71d: On the venous phase, the splenic vein is not visualized and the splenic outflow is seen to drain towards gastric veins and esophageal varices (△).



71e

7Ie: Splenic artery embolization with 3 steel coils is then performed (19–2–82). Two coils are in the intrasplenic branches and the third one in the main stem of the splenic artery. The patient remains well since that time and no variceal bleeding has recurred for 21 months.

71f



7If: Angiographic checking 21 months later (28–6–84). The spleen size is only slightly reduced (17 cm) and the splenic artery remains patent in spite of the previous embolization.
The steel coil within the splenic arterial trunk results only in a minimal narrowing (↑) without impairment of the splenic blood flow.
The splenic vein is well opacified whereas it was not visualized on the previous arteriography and the hepatic artery is a little less widened.
Therefore it is likely for the relatively good result in this patient to be due mainly to an improvement of the intrahepatic obstruction.

8. SEGMENTAL INTRAHEPATIC OBSTRUCTIONS WITHOUT PORTAL HYPERTENSION

Intrahepatic lesions involving a lobe or one or several segments of the liver may sometimes determine a compression of the surrounding hepatic venous bed severe enough to result in an interruption of the portal inflow in the corresponding area. Under these circumstances, there is no portal hypertension since a normal portal flow persists in the other hepatic territories.

The above-mentioned segmental intrahepatic obstructions have no consequences from a hemodynamic point of view and would remain unknown if angiographic methods did not demonstrate their existence due to the reversed intrahepatic portal flow resulting from them. Indeed, owing to the very low pressure gradient which normally exists between the portal system and the hepatic venous system, an increased sinusoidal pressure rapidly results in a slowed then interrupted portal flow in the corresponding area.

As a result, hepatic arterial flow increases to compensate for the loss of portal flow and intrahepatic arterio-portal communications develop allowing the arterial blood to flow back through the portal veins as soon as the obstruction is tight enough.

Once the reversed portal blood flow reaches a collateral portal vein feeding a normal hepatic territory where the portal inflow is normal, it drains through this portal branch in the hepatopetal direction.

This segmental reversal of the intrahepatic portal flow is an interesting phenomenon not only from a physiopathological point of view but also as far as diagnosis is concerned, for three reasons:

- It gives rise on arterioportography as well as on dynamic C.T. to abnormalities which might be wrongly attributed to morphologic changes.
- It should not be misinterpreted as an intrahepatic arterioportal fistula, the angiographic pattern and the causes of which are different.
- In some cases it may represent the one angiographic sign allowing for an intrahepatic focal hypovascular lesion to be diagnosed.

I. Segmental reversal of the intrahepatic portal flow: angiographic pattern

Although involving only a segment or a lobe of the liver the angiographic pattern of the segmental reversal of the intrahepatic portal flow is exactly the same as in cirrhosis (Chapter 5).

1. Direct sign

The direct sign is an early retrograde opacification of the intrahepatic portal veins in the involved area on selective hepatic arteriography. Unlike intrahepatic fistulae, this arterio-portal shunting is relatively delayed, occurring not immediately but 4 to 6 seconds after the injection of contrast medium into the hepatic artery.

It involves several small intrahepatic portal branches but the arteriovenous communications proper are too small and too distally located to be visualized. Furthermore, visualization of the smallest intrahepatic portal branches is in most cases hindered by a dense arterial hepatography so that only segmental portal branches are clearly evidenced.

Retrograde filling of the intrahepatic portal braches is all the better as the blood flow rate through the arterio-portal shunts is higher, which probably depends on the degree and location of the intrahepatic venous obstruction. In some cases, retrograde portal opacification involves one liver lobe completely and the contrast medium is seen to reach the portal bifurcation (Fig. 77), whereas in other cases it is restricted to a few small and distal portal branches (Figs. 78, 81–86).

2. Indirect signs

Two indirect signs may be helpful for the diagnosis: The first one is the demonstration of an arterial

hypervascularisation in the involved area on hepatic arteriography. Segmental arterial branches are dilated. This is mainly evidenced at their distal ends which are widened, with unsharp margins and sometimes duplicated which might correspond to an early opacification of some portal venules (Figs. 72, 81, 85). This distal hypervascular pattern associated with an increased arterial hepatography in the corresponding area is usually easy to recognize as compared to the normal neighbouring territories.

The second indirect sign is of course the lack of opacification of the intrahepatic portal branches in which the blood flow is reversed on the venous phase of superior mesenteric arteriography (Fig. 72c).

This functional obstruction pattern at the site where the two opposite blood flows meet is associated with a lack of portal hepatography in the same area.

Unlike a true obstruction the interrupted portal vein ends unsharply because the contrast medium is progressively diluted by the reversed non-opacified portal blood flow.

The functional origin of this portal venous obstruction is of course confirmed by the retrograde filling of the lacking portal branches through selective hepatic arteriography (Figs. 77, 78, 82, 83, 85).

This segmental reversal of the intrahepatic portal flow is then easily documented by arterioportography. However, it can be evidenced through dynamic C.T. using intravenous bolus injection of contrast medium as already demonstrated in cirrhosis (Fig. 45) and Budd-Chiari syndrome (Fig. 54).

This hemodynamic abnormality, once demonstrated, raises two main problems: 1) it must not be misinterpreted as an intrahepatic arterioportal fistula, and 2) it results from various hepatic lesions which must be recognized.

II. Differential diagnosis of segmental reversal of intrahepatic portal flow: arterio-portal fistulae

Segmental reversal of the intrahepatic portal flow seems to have been wrongly interpreted as an arterio-portal fistula in many cases, as both abnormalities are often described as 'hepatic arterioportal shunt'. They are, in fact, two different phenomena whose angiographic pattern and causes are different.

An intrahepatic arterioportal fistula is usually a unique and relatively proximal arterioportal shunt due to a lesion of the hepatic parenchyma, resulting in an abnormal tract between an arterial branch and the corresponding portal vein.

On hepatic arteriography the very early opacification of an intrahepatic portal vein, 1 to 2 seconds after the injection of contrast medium, is typical of an arterio-portal fistula. Since the shunt is usually proximally located on segmental or subsegmental vessels, the communication itself is clearly visible (Fig. 73).

This angiographic pattern is quite different from that of the reversal of the intrahepatic portal flow due to a post-sinusoidal obstruction. On the contrary, the same functional obstruction of the portal branches involved by the shunt is seen in both cases on the venous phase of S.M.A.

However, unlike the reversal of the intrahepatic portal flow, the portal vein opacified through a fistula fills in both hepatopetal and hepatofugal directions on hepatic arteriography, which is some times better appreciated by dynamic C.T. As a matter of fact, dynamic C.T. using intravenous bolus injection of contrast medium permits these arterio-portal fistulae to be demonstrated (156, 157, 158, 161) through an early opacification of an intrahepatic portal vein at the arterial phase of the injection (Fig. 76). Arterioportography as well as C.T. can also show other abnormalities associated with the cause of the fistula which is usually easily recognized.

1. Traumatic arterio-portal fistulae

Injury to the liver parenchyma either due to trauma or iatrogenic, resulting from percutaneous biopsy, portography or cholangiography is the main cause of intrahepatic arterio-portal fistulae (158, 159).

The arteriographic pattern is that of an early, unique and relatively proximal shunt, without any associated change, easy to differentiate from the segmental reversal of the intrahepatic portal flow (Fig. 73).

On dynamic C.T., direct visualization of the shunt at the arterial phase of the injection is associ-

ated with a dense opacification of a triangle-shaped area of the hepatic parenchyma due to the increased blood flow through the fistula.

It signifies that the sinusoidal filling from the arterial route is increased, since the hepatopetal blood flow through the normal liver parenchyma is preserved in case of fistula (158).

2. Tumoral arterio-portal fistulae

Arterio-portal fistulae are well known to occur in cases of primary liver carcinoma. They result from involvement of the portal venous system by tumoral growth, and their angiographic pattern was well described in many reports.

Tumoral arterio-portal fistulae are easily recognized as they are always associated with a large hypervascular tumor within the liver parenchyma. They are also well documented through dynamic C.T., as already demonstrated (157, 160).

3. Congenital arterio-portal fistulae

Hemangiomas, arterio-venous aneurysms and arterial aneurysms ruptured into the portal system can be responsible for an intrahepatic arterio-portal fistula but these congenital vascular lesions are in fact very rare.

4. Arterio-portal fistula, cirrhosis and portal vein thrombosis

Some rare cases of arterio-portal fistula associated with cirrhosis and portal vein obstruction were previously reported (162, 163, 164, 165). As a matter of fact, these two diseases are known to be favorable conditions for the development of intrahepatic arterioportal shunts.

In the case of portal vein thrombosis such shunts may result from the sudden drop of portal pressure in the patent intrahepatic portal veins located downstream the thrombus (Chapter IV), while in cirrhosis they are induced by the specific pathologic changes of the liver parenchyma (Chapter V). The association of both conditions might therefore be expected to give rise to more important arterio-portal communications than usual.

However, although cirrhosis and portal vein thrombosis can be associated with arterio-portal shunting resulting in the reversal of the intrahepatic portal flow, they are not known to be responsible for the development of true arterioportal fistulae.

Bookstein et al. reported seven cases of cirrhosis associated with a portal obstruction due to thrombosis in one and end-to-side porto-caval anastomosis in six. Indeed, in most of these cases the angiographic pattern of the arterioportal shunting was that of the reversal of the intrahepatic portal flow (162). However, in two out of the seven cases the shunt was proximally located and the portal vein filled very early in a hepatopetal direction so that the angiographic pattern was that of a fistula.

A large arterioportal fistula associated with a portal vein thrombosis was reported by McLoughlin et al. in a case of a cholangiocarcinoma of the liver complicating hemochromatosis (163). This fistula was shown to be due to the portal vein thrombosis and not to the liver carcinoma, which was an hypovascular tumor.

In the case reported by Sniderman et al., an arterioportal fistula resulted from hepatic schistosomiasis associated with a post-sinusoidal obstruction and portal vein thrombosis (164).

Lastly we reported a case of alcoholic cirrhosis associated with portal vein thrombosis in which an arterioportal fistula involving the left portal branch was evidenced through arteriography. This patient who had undergone neither liver biopsy nor percutaneous transhepatic radiological exploration died rapidly from hepatic failure.

The diagnosis was confirmed by autopsy and no other lesion than micro-nodular atrophic cirrhosis which might have accounted for the arterio-portal fistula was found through careful anatomical examination of the liver. In particular, a liver carcinoma associated with the cirrhosis could be excluded (165).

It is interesting to note that in that patient hepatic arteriography evidenced not only a large arterioportal fistula proximally located in the left lobe, but also a few smaller, more distally located and later visualized arterioportal shunts in the right lobe resulting in the typical angiographic pattern of the reversal of the intrahepatic portal flow (Fig. 74).

It is anticipated that these rare cases of arterioportal fistulae reported in cirrhosis associated with a portal vein obstruction might be due to an overdevelopment of one of the numerous and usually smaller and more distally located arterioportal communications which are well known to occur commonly in cirrhosis and result in reversed intrahepatic portal flow. As a matter of fact, one can expect a portal vein thrombosis to be responsible for an increased flood flow rate through any preexisting arterioportal communication since it results in a decreased venous pressure in the intrahepatic portal venous bed beyond the obstruction.

It is then suggested that various types of arterioportal shunts might occur in cirrhosis, resulting in either the angiographic pattern of the reversal of the intrahepatic portal flow or that of a true fistula. In other words, arterioportal communications are either multiple, distally located too small to be visualized, or more proximally located, and wide enough to be clearly visible, likening a true fistula.

The occurrence of either type of arterioportal shunt might depend on either hemodynamical factors such as those resulting from portal vein thrombosis for instance or anatomical ones such as specific pathological changes of the liver parenchyma related to the various types of cirrhosis or hepatic fibrosis.

In some cases an intermediate angiographic pattern between that of the reversed intrahepatic portal flow and an arterio-portal fistula may be encountered: the arteriovenous communications involve the segmental vessels and are visualized two seconds after the beginning of the injection of contrast medium in the hepatic artery. As a result, most of the segmental portal veins opacified in a retrograde direction are visible at the arterial phase alongside the corresponding arteries (Fig. 75).

Two such cases have been encountered in nonalcoholic patients in whom a diffuse intrahepatic obstruction was associated with a pronounced atrophy of the liver. One of them had a periportal fibrosis from unknown origin associated with portal vein thrombosis and responsible for an intrahepatic presinusoidal obstacle as hepatic venography and wedged hepatic vein pressure were normal (Fig. 75).

Owing to the portal vein thrombosis, the contrast medium filling the intrahepatic portal branches in a retrograde direction through the arterioportal shunts was seen to flow in a hepatofugal way through collateral pathways running into the hepatic pedicle as was reported by Reuter and Novak (55, 56) following an end-to-side portocaval anastomosis (Chapter 7, 2-2) (Fig. 75).

It is then demonstrated that the development of intrahepatic arterio-portal shunts leading to a reversed portal flow can occur not only in the case of post-sinusoidal obstruction but in the case of intrahepatic pre-sinusoidal obstruction as well. It is however, a surprising and probably very uncommon situation which implies that the arterio-portal shunts need to be much more proximally located than in cirrhosis since they are of course situated upstream the pre-sinusoidal obstacle. In other words, this curious hemodynamical condition implies that an obstruction involving the distal end of the intrahepatic portal veins between the arterioportal shunts and the normal sinusoids does exist.

This type of arterioportal communication that developed within a periportal sclerosis probably results from the specific pathological changes related to this peculiar disease although the portal vein thrombosis can also play a role in their occurrence.

Finally, although the reversal of the intrahepatic portal flow and the arterioportal fistulae usually corresponds to different lesions on the basis of their angiographic pattern as well as from a physiopathological and etiological point of view, both types of arterio-portal shunts and intermediate forms can probably occur in some cases of cirrhosis or periportal fibrosis.

The multiple small and distal arterio-portal shunts responsible for the angiographic pattern described as the reversal of the intrahepatic portal flow are by far the commonest.

The wider, more proximal, early opacified arterio-portal shunts, likening a true fistula may probably occur in some rare cases depending on either hemodynamical factors such as those resulting from portal vein thrombosis or anatomical factors such as fibrosis and atrophy of the liver parenchyma even in the case of a presinusoidal obstacle.

5. Arterio-portal fistula and cirrhosis

As arterioportal shunting is a common finding in cirrhosis and since the shunts may sometimes have the angiographic pattern of a true fistula when associated with portal vein thrombosis, one may wonder whether the cirrhotic pathologic changes themselves can give rise to a true intrahepatic arterioportal fistula, even though the portal vein remains patent. As a matter of fact, the association of a cirrhosis with an intrahepatic arterioportal fistula is a rare condition, which raises interesting questions from a physiopathologic as well as from a diagnostic point of view.

When portal hypertension is not known to be related to cirrhosis because either the clininical background is misleading or the pathological changes are considered not specific enough on liver biopsy, the arterio-portal fistula may be wrongly interpreted as responsible for portal hypertension. However, the possibility for an arterioportal fistula to give rise to portal hypertension is questionable (Chapter VI), and it seems difficult to anticipate that a liver cirrhosis could result from such a fistula.

Two other possibilities have then to be discussed: either there is no relationship whatsoever between cirrhosis and fistula or the intrahepatic arterioportal fistula results from the cirrhotic changes of the liver parenchyma.

Although this question cannot be clearly answered the previously reported cases enable one to estimate the second hypothesis as a possible one. Indeed, it is conceivable that a pronounced atrophy of a liver segment associated with a marked destruction and fibrous replacement of the hepatic lobules may all the more induce the development of intrahepatic arterio-portal shunts in this segment as atrophy of the liver parenchyma is more important.

In a patient known to have had a liver cirrhosis probably due to alcohol for eight years and referred for recurrent bleeding from esophageal varices, an arterioportal fistula was evidenced through arteriography in segment 8 of the right liver lobe (Fig. 76). The fistula was also visualized through dynamic C.T., which demonstrated an unusually pronounced shrinking of this segment (Fig. 76B).

This patient, of course had undergone neither liver biopsy nor percutaneous transhepatic exploration in the past and the diagnosis of cirrhosis was confirmed through surgical exploration.

Therefore, it is suggested that in cirrhosis a pronounced segmental atrophy might induce the development of a true arterioportal fistula within this area. This hypothesis is all the more probable as abnormal arterio-portal shunts can occur in case of segmental atrophy of the liver in non-cirrhotic patients as discussed later (Chapter 7, 4-4).

III. Etiological diagnosis

Segmental reversal of the intrahepatic portal flow is a hemodynamical phenomenon resulting from any obstruction of the hepatic venous bed whatever its site and its cause. It is mainly encountered in hepatic tumors, liver trauma and some cases of extrahepatic masses as well (166).

1. Hepatic tumors

Involvement of the hepatic veins running in close contact to a hepatic tumor can give rise to a segmental post-sinusoidal obstruction.

When the main hepatic veins are obstructed in their suprahepatic part this results in a secondary Budd-Chiari syndrome from tumoral origin. However, the intrahepatic venous bed can be involved as well, while the main stems of the hepatic veins remain patent, which seems to be a more common condition (167). Various tumors can be responsible for this phenomenon such as primary liver carcinoma, hepatic metastases, as well as some cases of benign tumors and subcapsular hematoma of the liver.

a) Primary carcinoma of the liver

Reversal of the intrahepatic portal flow is essentially associated with hepatic tumors demonstrating a hypovascular and infiltrating pattern from a pathological point of view. Conversely, hypervascular hepatomas can give rise to a tumoral arterioportal fistula so that even though the hepatic veins are involved by the tumor the subsequent reversed portal flow will be either hidden by the fistula or confused with it.

In practice, when arterioportal shunting is associated with a hypervascular malignant tumor of the liver the question as to whether abnormal opacification of the portal veins results from a reversal of the portal flow or a tumoral fistula cannot be determined. On the contrary, when the carcinoma is a hypovascular tumor as in cholangiocarcinoma and some cases of hepatoma, there is no possibility for a tumoral arterioportal fistula to develop. Hence, when arterioportal shunting occurs under this circumstance it always results from a tumoral involvement of the hepatic venous bed which can be documented through hepatic venography (Fig. 77), and its angiographic pattern is that of the segmental reversal of the intrahepatic portal flow.

This hemodynamic phenomenon has proved to be a useful diagnostic criterion when occurring in infiltrating and poorly delineated malignant tumors, the margins of which are not outlined by distortion and displacement of the surrounding vessels, a common case in hepatic masses.

It is then possible for the reversed portal flow in the hepatic area involved by the neoplasm to be the main if not the one sign allowing for the tumor to be recognized as there is neither neovascularity nor the typical angiographic pattern of a mass (Fig. 77).

b) Metastases to the liver

Like the primary carcinoma, the hypovascular metastases can be responsible for the segmental reversal of the intrahepatic portal flow when they are either large-sized or diffuse enough to produce an obstruction of the hepatic venous bed within the liver.

Reversal of the portal flow is usually minimal, only a few small portal branches being visualized in a localized area (Fig. 78). It is associated with a moderate dilatation of the small distal arterial branches and increased arterial hepatography in the liver parenchyma surrounding the metastases.

This moderate and distal hypervascular arterial pattern which was sometimes reported as a common angiographic sign of hypovascular metastases to the liver is then probably an indirect sign of the reversed portal flow, i.e. it means that the arterial flow increases to compensate for the lost portal flow resulting from compression and obstruction of the sinusoids and the hepatic venules by the metastases.

Indeed, this mild dilatation of the distal hepatic arterial network seems to be a common angiographic finding in diffuse hypovascular hepatic metastases whereas the retrograde opacification of the portal veins in the corresponding area is rather rare. This might be due to the fact that either the post-sinusoidal obstruction is not severe enough for the hepatopetal portal flow to be completely interrupted or the portal venules are narrowed together with the hepatic veins in the involved area.

However, the same distal arterial hypervascular pattern is always demonstrated wherever the intrahepatic portal flow is reversed, whatever the causative factor and especially in case of benign tumor and liver trauma. Therefore, it is not likely to represent a tumoral neovascularity, contrary to what might be thought, but a hemodynamic phenomenon related to a decreased or interrupted portal flow in the corresponding area.

The increased arterial hepatography around the tumoral lesions usually appears as patchy areas of increased density but can also sharply outline the tumors giving rise to the pattern of ring enhancement. This ring enhancement surrounding an intrahepatic mass was formerly demonstrated by arteriography in hydatic cysts of the liver. It is, however, a non-specific sign which can be associated with any intrahepatic avascular mass such as biliary cyst, benign tumors and metastases to the liver as well.

The precise anatomical significance of this rim enhancement pattern is not clearly known although a few different explanations were proposed. Owing to its possible association with a reversed portal flow it may then be suggested to represent a hemodynamic phenomenon related to an increased arterial flow in the normal parenchyma surrounding the tumor. As a matter of fact, the compression of the normal hepatic parenchyma surrounding an intrahepatic mass results in an impairment of the normal blood flow through the corresponding sinusoid capillaries and small hepatic venules.

An interruption of the hepatopetal portal flow in the thin layer of normal hepatic parenchyma surrounding the mass could then result in an increased arterial flow with stagnation of the contrast medium in the compressed area giving rise to this peculiar angiographic pattern.

This ring enhancement is nowadays commonly

demonstrated through dynamic C.T. around the hepatic metastases (168, 169).

Moss et al., by comparing the results of angio C.T. with either intravenous or intra-arterial injection of contrast medium, demonstrated that the hepatic arterial blood flow is responsible for this pattern as the ring enhancement in metastatic lesions was not as well defined after intravenous bolus injection as after intraarterial bolus (168). The authors conclude that it is probably due to the fact that most hepatic lesions receive their blood supply from the hepatic arterial circulation. However, the hemodynamic explanation seems more likely as the same pattern can be encountered in liver cysts as well as in metastases. Furthermore, this ring shadow was reported to be associated with areas of increased density occurring at the arterial phase of dynamic C.T. in the normal hepatic parenchyma surrounding the tumor (169). Indeed it is not an uncommon C.T. finding in either primary or secondary hepatic neoplasms (Figs. 79 & 80).

Itai et al. anticipated that these areas of increased density surrounding the hepatic tumors could be due to an increased hepatic arterial flow resulting from tumoral involvement of the portal vein (169). However, it is an inconstant finding in hepatic carcinoma associated with portal vein thrombosis while it may be also evidenced in case the portal vein is normally patent. It seems therefore more likely that this increased arterial flow as demonstrated through arteriography, together with dynamic C.T., is due to decreased portal inflow resulting from an obstruction of the hepatic venous drainage of the normal liver parenchyma surrounding the tumor.

Finally, it is suggested that the arterial hypervascular pattern of the normal hepatic parenchyma surrounding a hypovascular tumor of the liver is a hemodynamic abnormality resulting from tumoral involvement of the hepatic venous bed. Its significance is then the same as the segmental reversal of the intrahepatic portal flow while it is a much more frequent finding.

c) Benign hepatic tumors and cysts

Benign tumors and cysts are probably much more rarely responsible for a reversed intrahepatic portal flow than malignant tumors and yet this phenomenon remains minimal, resulting only in a faint retrograde opacification of some small distal portal branches. However, this angiographic finding was seen to be associated with three cases of hydatic cyst of the right liver lobe. In one of these cases, compression of the right hepatic vein by the posterior wall of the cyst could be demonstrated through hepatic venography as well as surgery (Fig. 81).

d) Sub-capsular hematoma of the liver

A reversed intrahepatic portal flow appears to be a common finding in the case of either traumatic or spontaneous subcapsular hematoma of the liver (170). Indeed, this blood collection is usually large and spreads from the anterior and lateral part of the liver to the posterior part at the level of the hepatic veins pedicle. It surrounds the whole right liver lobe, strongly compressing the hepatic parenchyma and the intrahepatic venous bed as well as probably the right hepatic vein, as demonstrated through hepatic venography (Fig. 82).

2. Extrahepatic masses

Some extrahepatic masses can give rise to a reversed intrahepatic portal flow when compressing the hepatic veins in their extrahepatic segment. It is, however, a rare condition which can result only from large posterior masses located behind the liver in the right upper abdomen.

Malignant tumors of the right kidney and of the right adrenal gland are known to involve the hepatic veins and give rise to a peculiar type of Budd-Chiari syndrome. This rare condition is in fact always associated with a thrombosis of the inferior vena cava. Indeed, some large, benign retroperitoneal masses can compress either the hepatic veins or the hepatic parenchyma probably in the same way as a subcapsular liver hematoma.

A reversed portal flow in the left lobe of the liver could be demonstrated in association with two such cases: a large pancreatic pseudocyst and a retroperitoneal cystic lymphangioma (Figs. 83, 84).

3. Liver trauma

Contusion of the liver is a common cause of the reversal of the intrahepatic portal flow, but this hemodynamic abnormality seems to have been misinterpreted in most cases as traumatic arteriovenous fistulae (171). Indeed, although traumatic arterio-portal fistulae do exist they are probably rare in liver contusions and easy to recognize in any case owing to their typical angiographic pattern.

On the contrary, reversal of the intrahepatic portal flow is more common and, unlike fistulae, it results from multiple, small, distal and usually invisible arterioportal communications, the opacification of which is relatively delayed.

These two different angiographic patterns of the arterio-portal shunts in liver trauma were formerly described by Boijsen, who noticed that the latter was more frequent and often a transient phenomenon (172).

According to a previous study, arterioportography performed within the first 48 hours following a contusion of the liver disclosed a reversed intrahepatic portal flow in 12 out of 26 patients (171). This angiographic finding is usually a transient abnormality except when it result from thrombosis of a relatively large hepatic venous branch within the liver as could be observed in one case of liver injury due to gunshot (Fig. 72).

Indeed, in most cases this reversal of the intrahepatic portal flow spontaneously disappears within 8 to 10 days, which correlates well with the fact that this arterioportal shunting is only a hemodynamic phenomenon which does not result from the development of traumatic arterioportal fistulae. It is usually associated with benign and spontaneously healing liver contusions.

Under these circumstances, hepatic venography, demonstrated the normal patency of the main stems of the hepatic veins but a retrograde opacification of the hepatic area where the portal flow was seen to be reversed, could never be obtained. Therefore, post-sinusoidal obstruction responsible for the reversed portal flow is likely to be located on the small hepatic venules and to be due to compression of the hepatic venous bed by the interstitial oedema and hematoma resulting from the contusion (Fig. 85). One can then understand that the reversed portal flow is a transient phenomenon as is the oedema, the spontaneous resorption of which allows the portal blood to flow again in the hepatopetal direction 8 to 10 days later.

This segmental reversal of the intrahepatic portal flow in liver trauma sometimes produces large defects in the portal hepatography, which resembles a mass lesion and might be misinterpreted as a severe organic lesion of the liver parenchyma although it is only a hemodynamic anomaly, which usually signifies a benign contusion.

4. Focal atrophy of the liver

Focal atrophic sclerosis of the liver parenchyma, whatever its cause, may also be responsible for a reversed portal flow in the corresponding area, as demonstrated in a case of chronic pancreatitis associated with a complete atrophy and a dense sclerosis of the left lobe of the liver (Fig. 86).

In that patient, the cause of atrophic sclerosis is unknown and one can only suggest that it could be a scarring process resulting from a previous involvement of the left lobe of the liver by pancreatitis. Indeed a large pancreatic pseudo-cyst proved to be able to compress the left lobe of the liver sufficiently to produce a reversal of the intrahepatic portal flow (Fig. 83). Furthermore, it is even possible for an acute bout of pancreatitis to produce necrosis of the hepatic parenchyma, resulting in a large hematoma within the left lobe of the liver, which compares similarly with the pancreatic pseudo-cyst occurring more commonly within the splenic parenchyma.

In any case surgery proved that the patient had a benign atrophic fibrosis of the left lobe of the liver, while the right lobe was normal and one can imagine that, when progressively evolving, such a fibrosis results in a decreased portal inflow compensated by an increased arterial flow and then in a reversed portal flow once the hepatic venous bed is completely obstructed by fibrosis and atrophy of the liver parenchyma.

This reversed portal flow associated with a focal atrophic sclerosis of the liver is probably a rare but interesting condition for two main reasons:

1) from a diagnostic point of view, the angiographic demonstration of such an arterioportal shunt with a reversed portal flow in a segment or a

lobe of the liver could be misinterpreted as suggesting a malignant tumor while it results only from a benign atrophic scar within the liver parenchyma.

2) from a physiopathological point of view, if arterioportal shunts can result from the development of atrophic sclerosis in a previously normal liver they should be more likely to occur when such a focal fibrosis and atrophy are associated with cirrhotic lesions.

For this reason one may wonder whether some intrahepatic arterio-portal fistulae reported in cirrhosis cannot result from an unusual overgrowth of some of the small common cirrhotic arterioportal communications owing to an unusually pronounced segmental atrophy of the liver (Fig. 76).

IV. Conclusions

- *Reversal of the portal flow in a localized area of the liver and aterio-portal fistulae are two different abnormalities* which must be clearly differentiated among the intrahepatic arterio-portal shunts evidenced through arteriography. Reversed intrahepatic portal flow corresponds to the development of normal intrahepatic arterio-portal communications resulting from a post-sinusoidal obstruction and associated with a hepatofugal portal flow.

  Conversely, an arterio-portal fistula is an abnormal communication between an intrahepatic artery and a portal vein branch, which is not associated with hepatic venous obstruction so that the intrahepatic portal flow remains normal.

  Although different from a physiological as well as angiographic point of view both phenomena can be associated in rare cases of cirrhosis or periportal fibrosis in which intermediate types of arterioportal shunts can also be encountered.
- Segmental reversal of the intrahepatic portal flow is a useful angiographic finding since it accounts for a post-sinusoidal obstruction whatever its cause and level.
- This angiographic or dynamic C.T. finding must not be misinterpreted as a morphologic lesion as it corresponds only to a hemodynamic abnormality.
- *The occurrence of this phenomenon in case of liver trauma, hepatic tumors and even some extrahepatic masses demonstrates that the intrahepatic arterio-portal communications are probably common in normal liver as they are easily evidenced as soon as a post-sinusoidal obstruction develops.*

72a

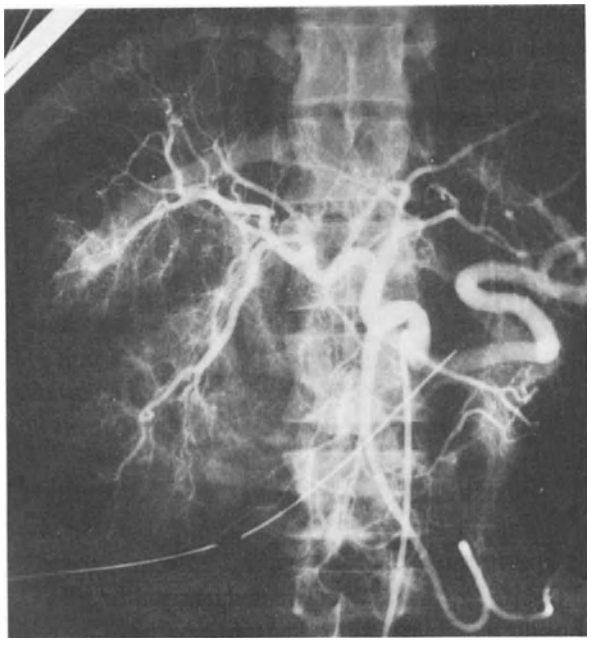
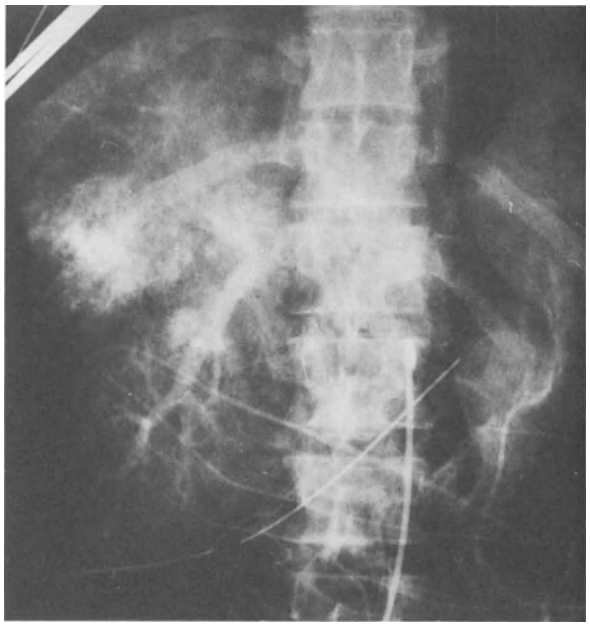
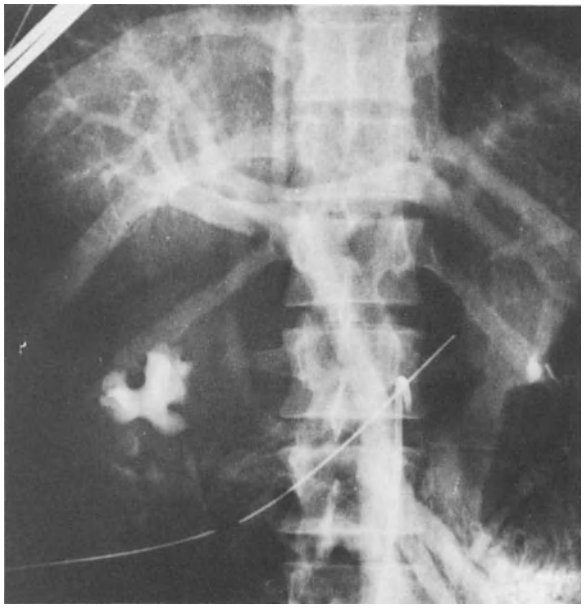


Fig. 72. Segmental reversal of the intra-hepatic portal flow resulting from liver trauma in a 24-year-old male.
9/11/74: emergency surgery following gunshot injury: large wound of the hepatic right lobe.
13/11/74: arterioportography.
72a: Coeliac arteriography:
Dilatation of the hepatic arterial branches in the segments 5 and 6 associated with a dense arterial hepatography in both areas.



72b

72b: 5 sec later, retrograde portal vein opacification is evidenced in the segment 5.



72c

72c: Venous phase of S.M.A.
Normal opacification of the portal veins in the left lobe and the upper right lobe but no filling of the portal branches corresponding to the lower right lobe (segments 5 and 6) where the flow is reversed.
6 months later a checking arteriography showed the same findings and a surgical biopsy proved the hepatic veins to be thrombosed within the liver parenchyma of the lower right lobe.

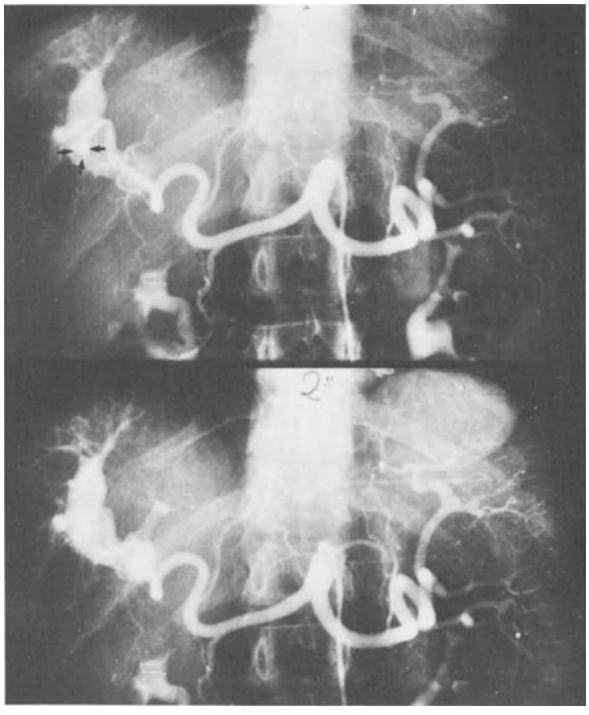
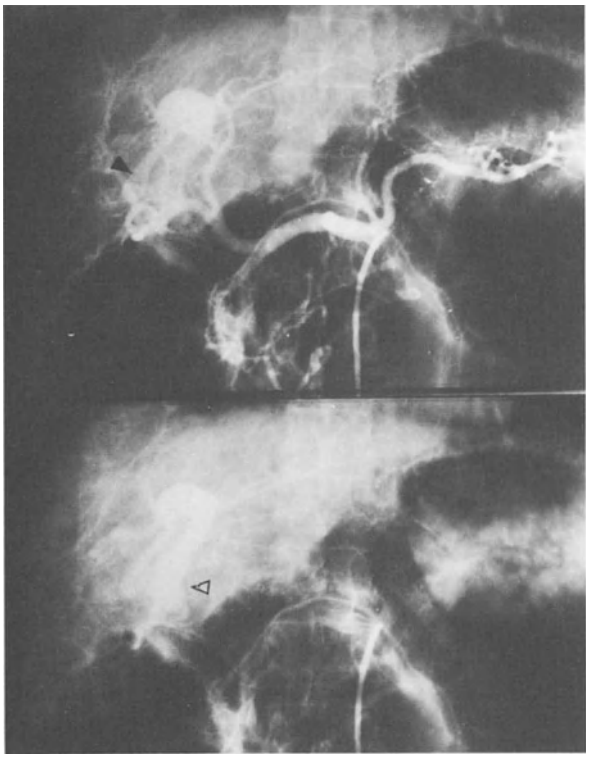
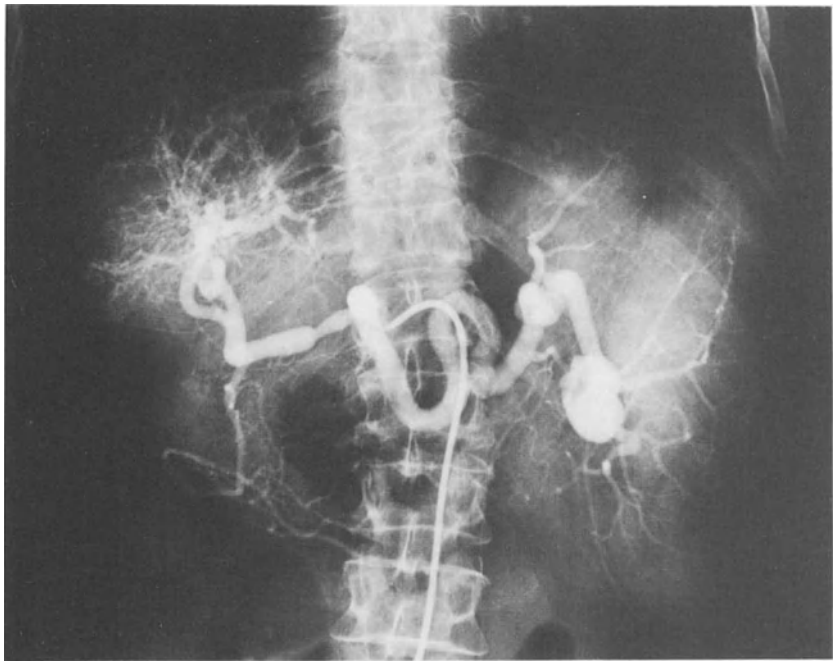


Fig. 73. Traumatic arterio-portal fistula resulting from a percutaneous liver biopsy performed 3 years earlier in a 41-year-old cirrhotic female.
Coeliac arteriography (13–9–76) demonstrates the typical angiographic pattern of an arterio-portal fistual: massive and early filling of an intrahepatic portal vein visualized at the first second following arterial injection of the contrast medium.
The abnormal arterio-portal communication is unique relatively proximal and can be identified (↑).
The flow in the portal branch opacified through the fistula is seen to be hepatopetal.

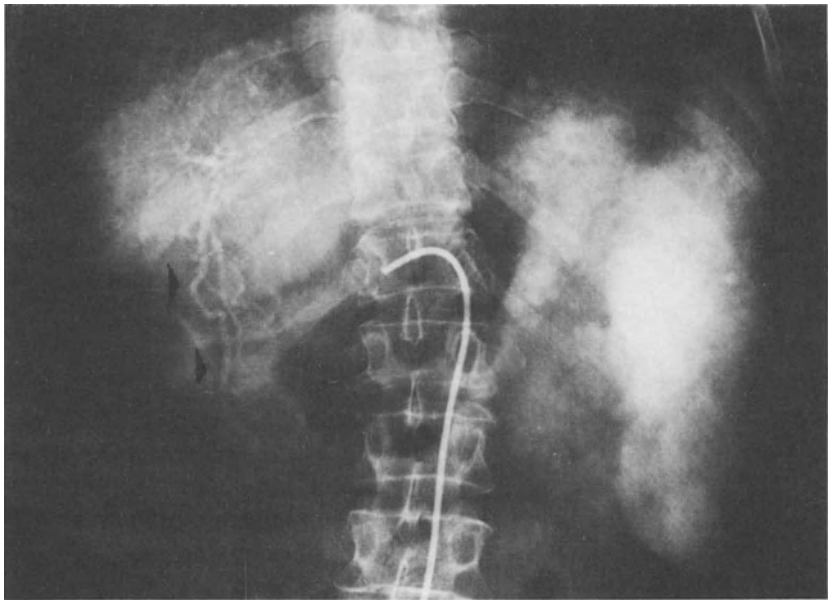
Fig. 74. Arterioportal fistula associated with alcoholic cirrhosis and portal vein thrombosis in a 62-year-old female.
Coeliac arteriography (30–1–75):
Massive and early filling (1st sec) of the left portal branch through a proximal arterio-portal communication located in the segment 4 (↑) (A).
Retrograde filling of the left portal vein is seen to reach the liver hilum but the portal stem is never visualized. (△) (B).
The splenic vein is not opacified on the late films and the superior mesenteric arteriography cannot be peformed because of thrombosis of the superior mesenteric artery.
The patient died from hepatic failure and anatomical exploration disclosed a complete thrombosis of the portal vein between the spleno-mesenteric confluence and the liver hilum.
The liver was atrophic and micro-nodular and no other lesion than cirrhosis was found within the liver parenchyma which could have been responsible for the fistula. Of course this patient never underwent any percutaneous puncture of the liver in the past.





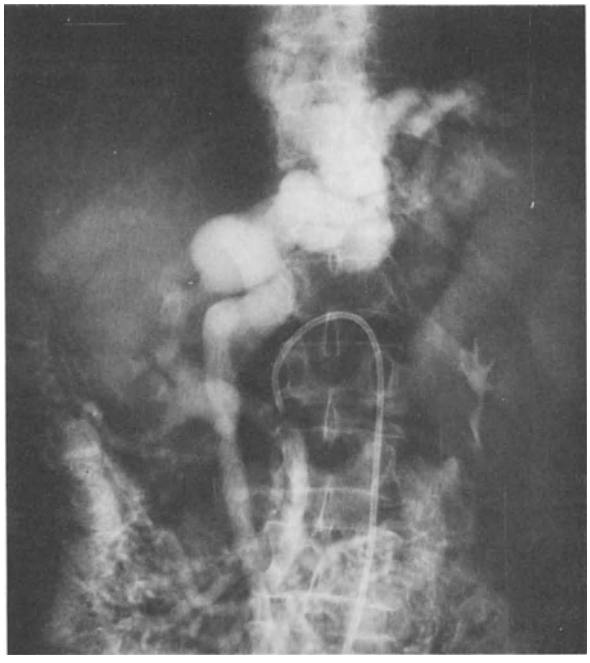
75a

Fig. 75. Esophageal varices discovered through endoscopy in a 62-year-old female referred for gastrointestinal bleeding thought to be due to peptic disease. No history of alcohol intake or liver disease in this previously healthy patient.
75a: Coeliac arteriography – 2nd sec (2–6–75):
Pronounced splenomegaly with splenic artery aneurysm accounting for a long-standing portal hypertension.
Marked and diffuse atrophy of the liver. Dilated hepatic artery (proximal spasm due to the catheter). Early retrograde filling of most of the small segmental portal branches running alongside the corresponding arterial branches.



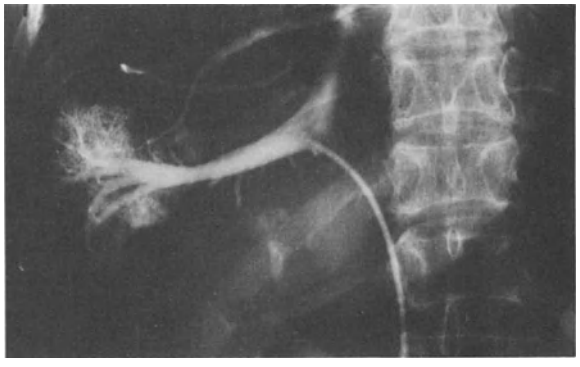
75b

75b: Coeliac arteriography – 6th sec (2–6–75):
The contrast medium filling the intrahepatic portal branches drains in a hepatofugal way through collateral venous pathways running into the hepatic pedicle (▲).



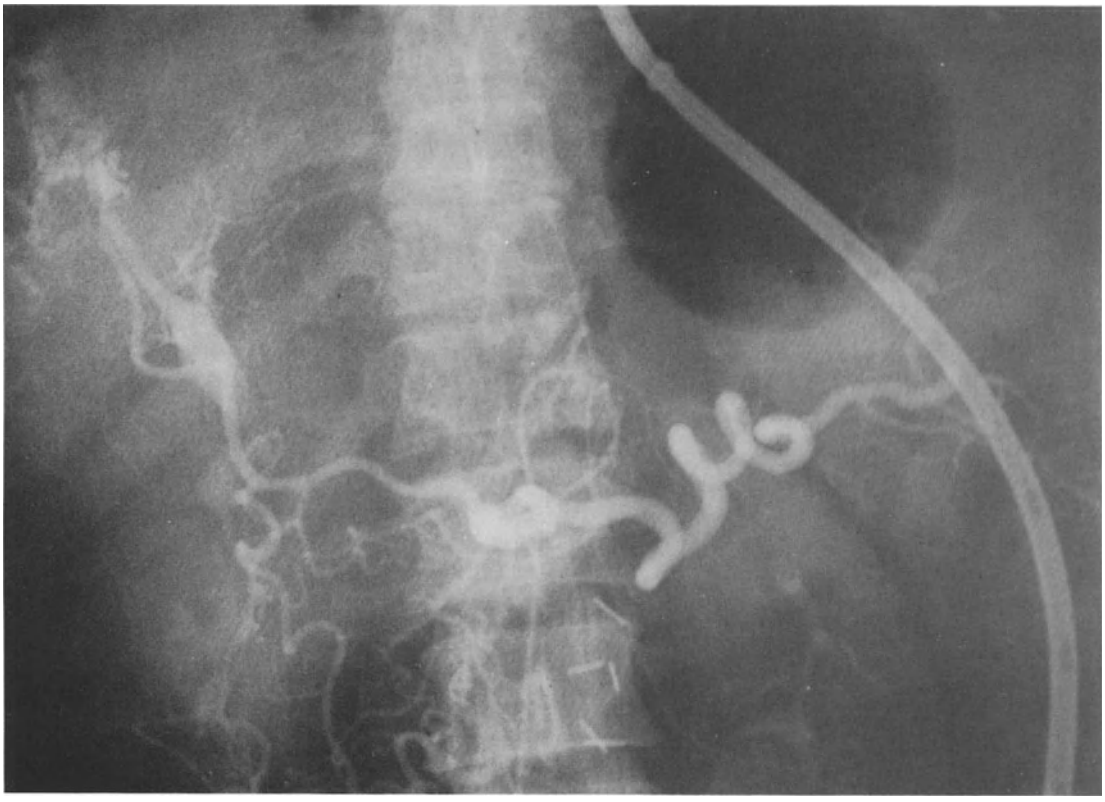
75c

75c: Venous phase of S.M.A. (2–6–75):
Complete obstruction of the portal vein with a completely hepatofugal portal flow through the left gastric vein and large gastro-esophageal varices. The lack of hepatopetal porto-portal collateral flow accounts for the existence of an intra-hepatic or suprahepatic obstacle associated with the portal vein thrombosis.



75d

75d: Hepatic venography) (17–6–75):
Normal hepatic veins and normal sinusoïdal filling. Normal wedged hepatic pressure: 11 cm saline.
A surgical porto-caval anastomosis is performed (15–9–75): 'Atrophic liver suggesting a post-necrotic cirrhosis.
Portal pressure: 20 cm saline. Irregular thickening of the wall of the superior mesenteric vein accounting for a long-standing endophlebitis'.
Liver biopsy: periportal sclerosis – steatosis-moderate annular sclerosis in some subcapsular areas.
Splenic artery biopsy: Pathologic changes likening a moderate fibro-muscular dysplasia.
In conclusion: hepatic fibrosis from unknown origin with intrahepatic presinusoïdal obstruction associated with portal vein thrombosis and intrahepatic arterioportal shunting.
The liver function tests abnormalities are only moderate. This patient is doing well seven years later.



76a

76b

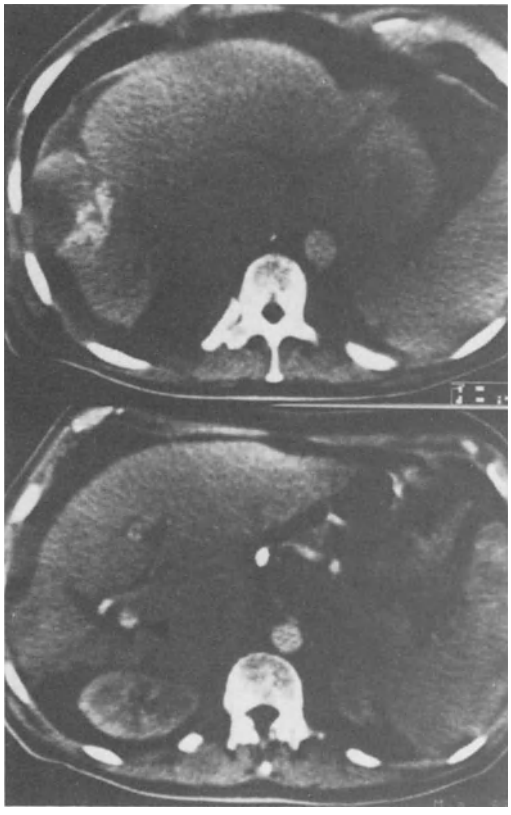
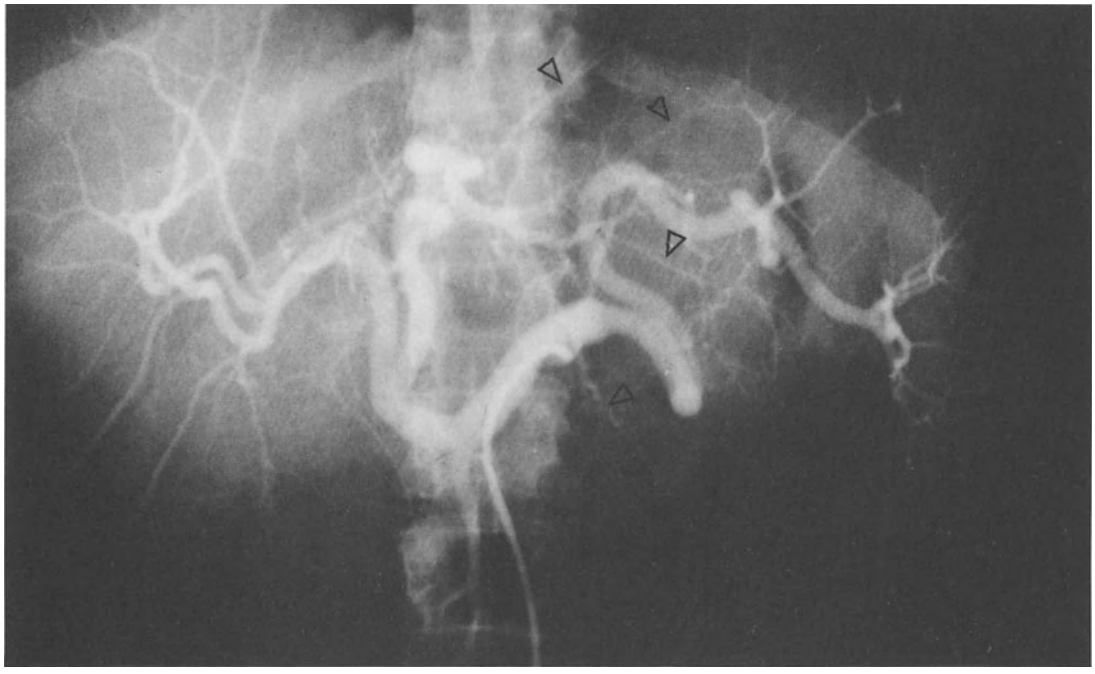


Fig. 76. Arterio-portal fistula in a 65-year-old male known to have cirrhosis from alcohol, for 8 years. First variceal bleeding in 1976.

76a: Arterioportography for recurrent bleeding (24–2–84): Early retrograde filling (2nd sec) of the portal branch of the segment 8 in a markedly atrophic right liver lobe through an arterio-portal fistula. After reaching the right portal branch the contrast medium was seen to drain through the other segments of the liver in the hepatopetal direction. The venous phase of S.M.A. showed the portal vein to be normally patent.

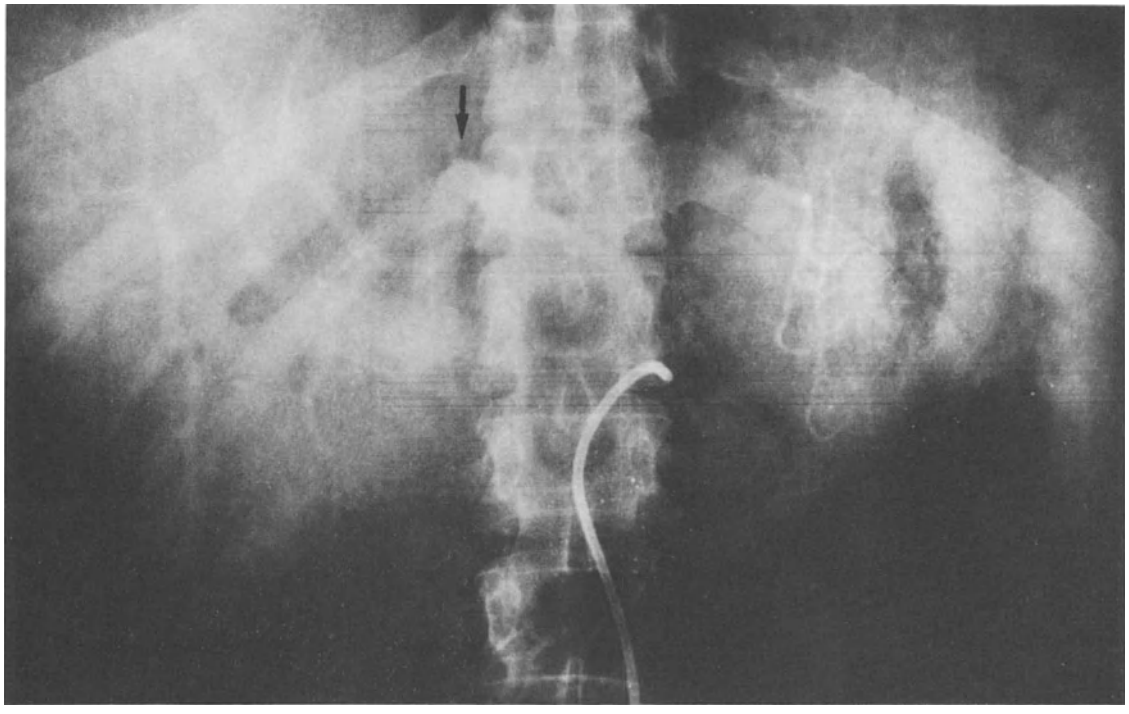
76b: The arterial phase of dynamic CT at two different levels visualizes the fistula (3–3–84). At the level of the hilum, early filling of the intrahepatic portal vein is seen alongside the corresponding arterial branch. Two centimeters higher the same vascular network as seen on arteriography is visualized.
In addition, a pronounced atrophy of the right liver lobe is evidenced while the left lobe is enlarged.
Atropho-hypertrophic cirrhosis was confirmed by surgery. Dilated vessels suggesting an angioma were seen around the atrophic right lobe.



77a

Fig. 77. Primary carcinoma of the left lobe of the liver responsible for a reversed portal flow in a 53-year-old male known to have hemochromatosis for 12 years.

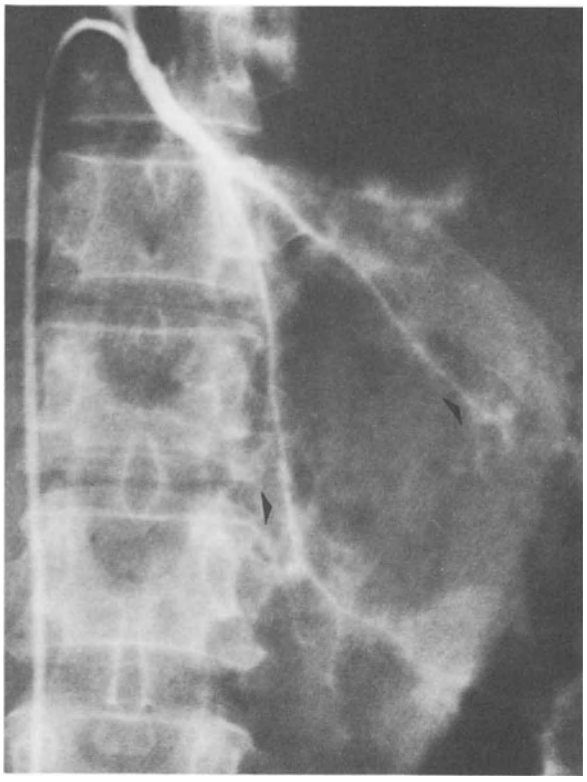
77a: Coeliac arteriography – 2 sec – (22–1–73).
Diffuse enlargement of the liver. The arterial branches within the left lobe are stretched and regularly spread apart from each other (△) but neither neo-vascularization nor a typical mass pattern is evidenced.



77b

77b: Coeliac arteriography – 5 sec – Retrograde filling of the left portal branch (↓) which drains towards the right one and then in the hepatopetal direction.
The right portal branch only was visualized through the venous phase of S.M.A. while the left one was not opacified.

77c



77c: Hepatic venography (6–2–73).
The left hepatic vein and its main intrahepatic branches are markedly narrowed (▲) while the main right hepatic venous branches are normally patent.
Cholangiocarcinoma of the left lobe of the liver is demonstrated through surgical exploration and biopsy.

78a

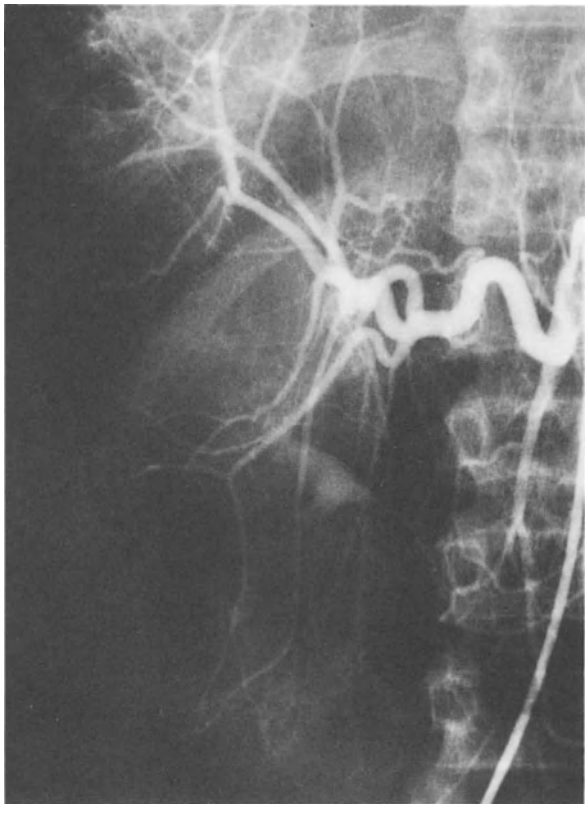
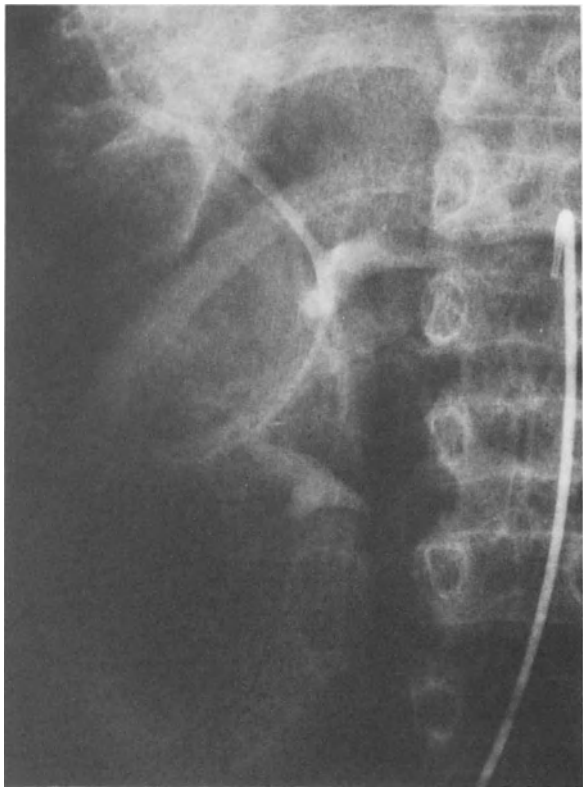


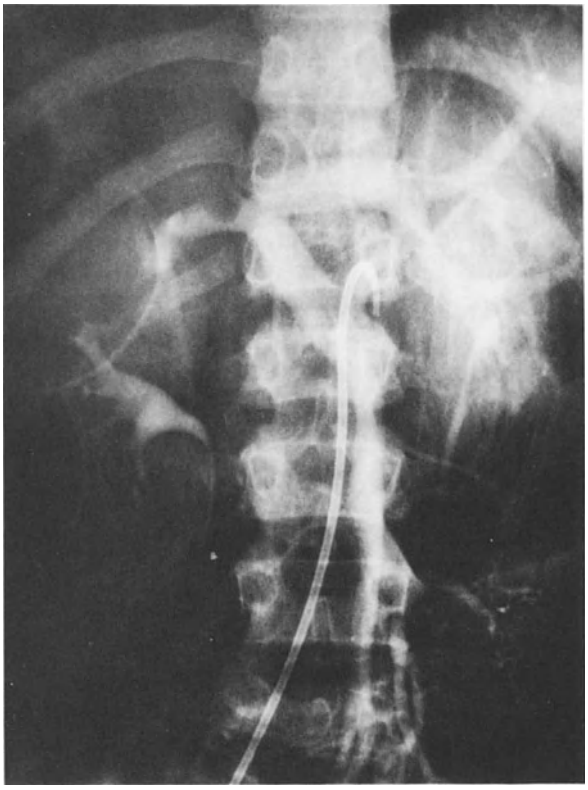
Fig. 78. Neuroblastoma revealed by metastases to the liver in a 13-year-old male.

78a: Selective hepatic arteriography – 3d sec – (4–6–76)
Enlargement of the right lobe of the liver Displacement of the arterial branches delineating several large hypovascular intrahepatic masses. Moderate dilatation of the distal arterial branches within the normal parenchyma surrounding the tumors at the upper and lower parts of the right lobe.



78b

78b: Selective hepatic arteriography – 6th sec – (4–6–76)
Retrograde filling of the intrahepatic portal branches surrounding the tumors with visualization of the right portal branch.



78c

78c: Venous phase of S.M.A. – (4–6–76)
Normal portal vein and left portal branch. Enlargement of the left hepatic lobe probably resulting from tumoral involvement of the right one. Functional obstruction of the right portal branches in which the flow is reversed. A defect in the right portal branch corresponds to either a thrombus or tumoral involvement but this vein remains patent.

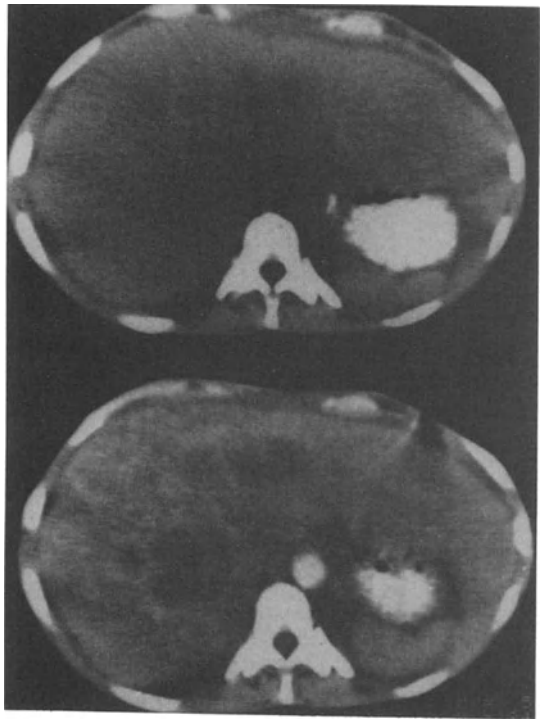


Fig. 79 Metastases to the liver originating from colon carcinoma in a 62-year-old female (20–9–82).
Plain C.T. (upper slice) shows multiple mass lesions of diminished density in both liver lobes.
Through dynamic C.T. (lower slice) an abnormally dense opacification of the hepatic parenchyma surrounding the metastases is evidenced during the arterial phase.
The pattern of ring enhancement surrounding most lesions is clearly demonstrated on the first slice following injection of the contrast medium.

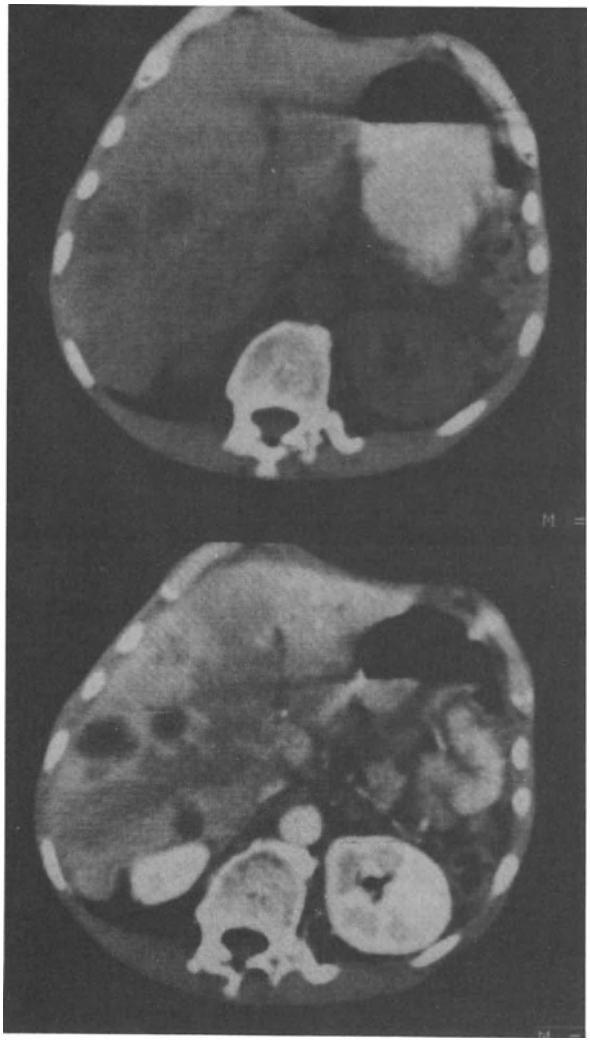
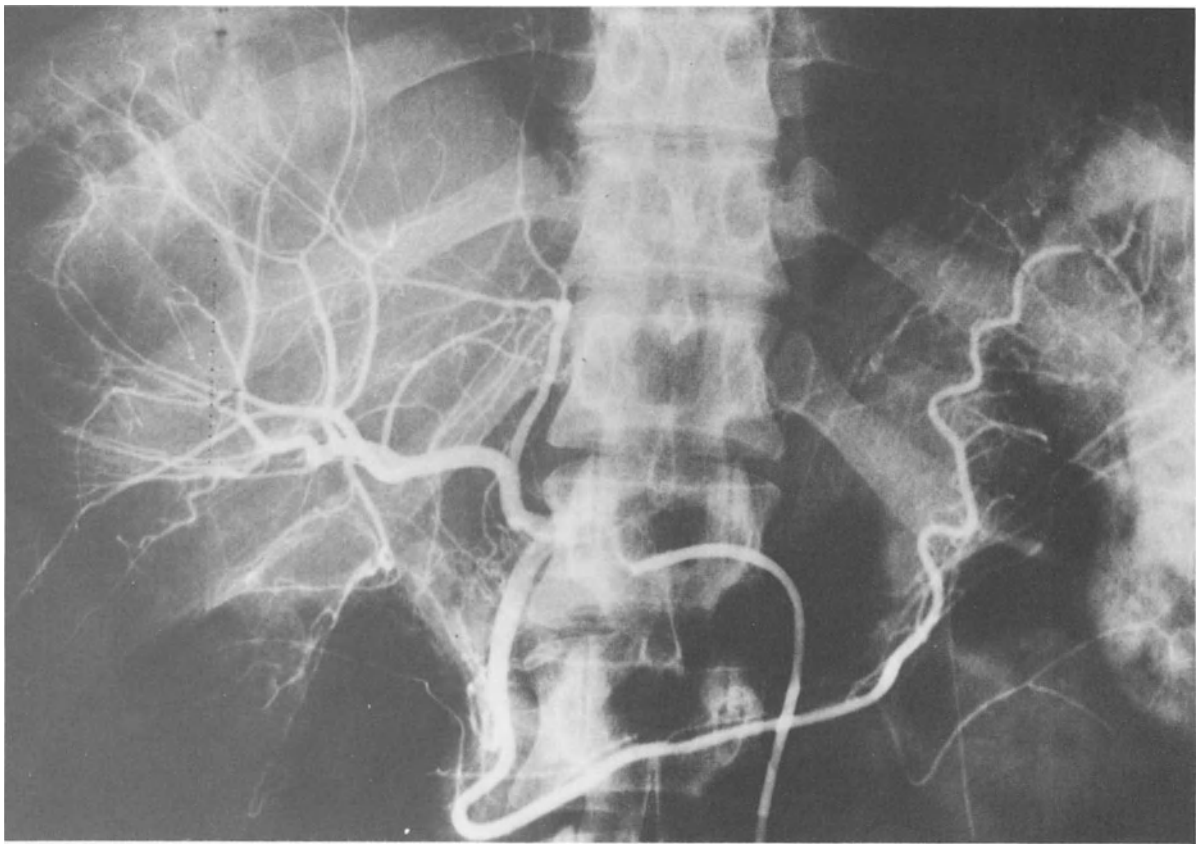
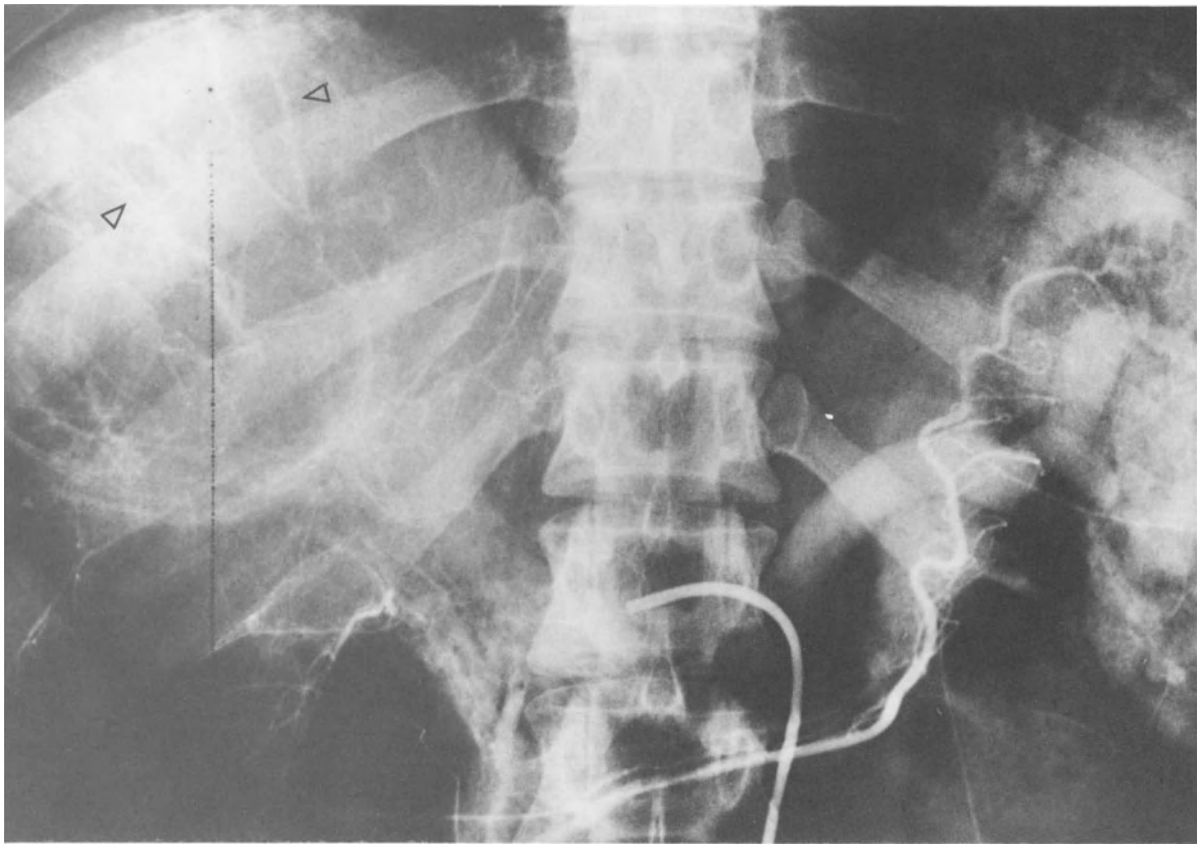


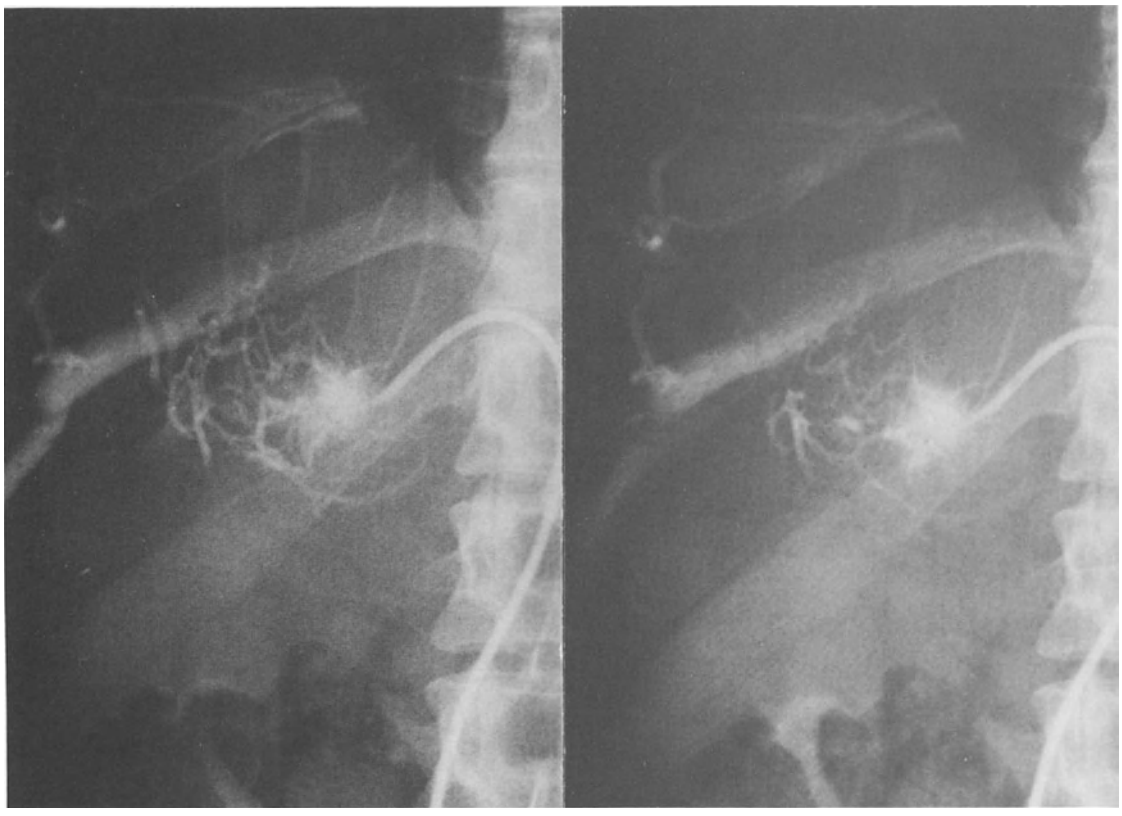
Fig. 80. Metastases to the liver originating from pancreatic carcinoma in a 52-year-old patient.
Several mass lesions of diminished density are demonstrated in the right lobe of the liver on plain C.T. (upper slice). Dynamic C.T. shows an abnormally dense opacification of the hepatic parenchyma surrounding the metastases during the arterial phase, accounting for an increased arterial flow in this area.



81a



81b



81c

Fig. 81. Large hydatic cyst involving the upper part of the right lobe of the liver in a 38-year-old patient.

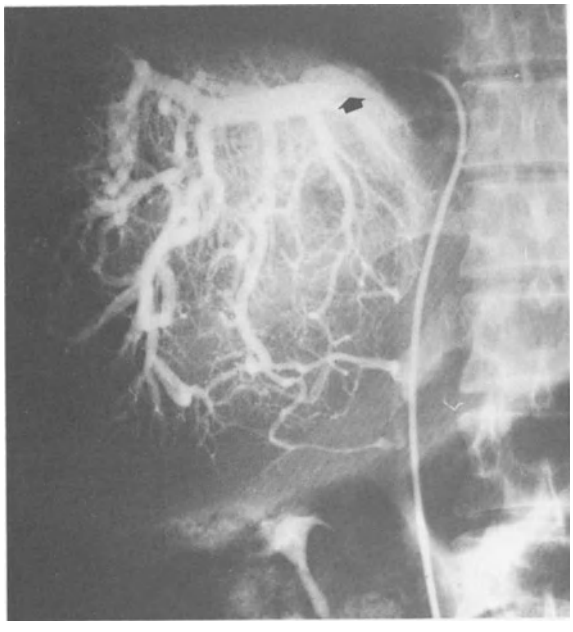
81a: Coeliac arteriography – 3rd sec – (12–6–74)
Displacement and stretching of the intrahepatic arterial branches around a large hypovascular mass.
At the upper pole of the mass some small arterial branches seem to be duplicated or surrounded by an unsharp stripe of contrast medium.

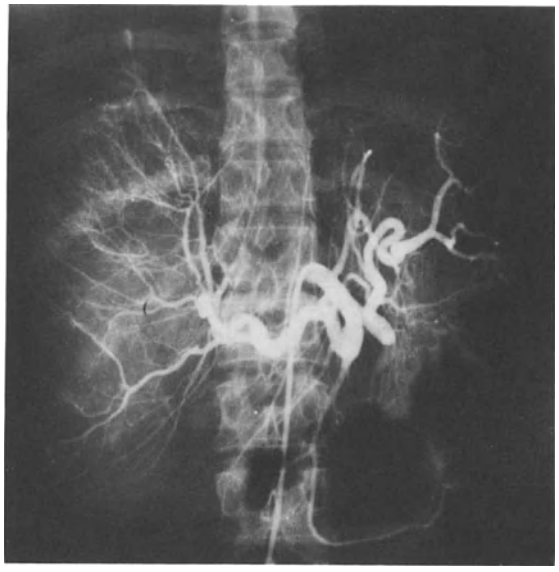
81b: Coeliac arteriography – 6th sec – (12–6–74)
This moderate and distal hypervascular pattern is clearly seen to correspond to retrograde filling of the small intrahepatic portal branches at the upper pole of the mass (△).

81c: Hepatic venography (16–6–74)
The main right hepatic vein cannot be catheterized. It opacifies through collateral ways following injection in a small accessory hepatic vein (left).
5 sec after the end of the injection, stagnation of contrast medium in the distal part of the right hepatic vein accounts for a compression of this vein (right).
At surgery (26–6–74) the posterior wall of the cyst is seen in close contact to the retrohepatic vena cava.

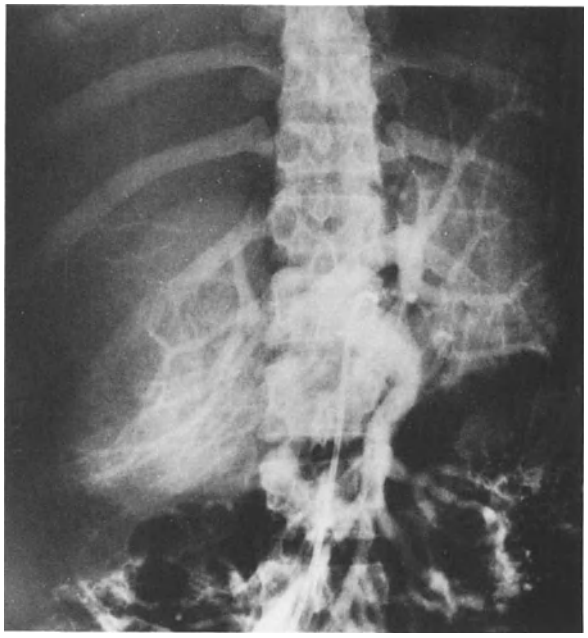
81d: Hepatic venography (24–5–83)
9 years later a checking angiography performed for jaundice shows the same focal reversal of the intrahepatic portal flow as previously documented. Hepatic venography demonstrates a persistent narrowing of the right hepatic vein (↑) which cannot be passed by the catheter tip and probably results from a fibrous scar due to the previous hydatic cyst.

81d





82a



82c

82b

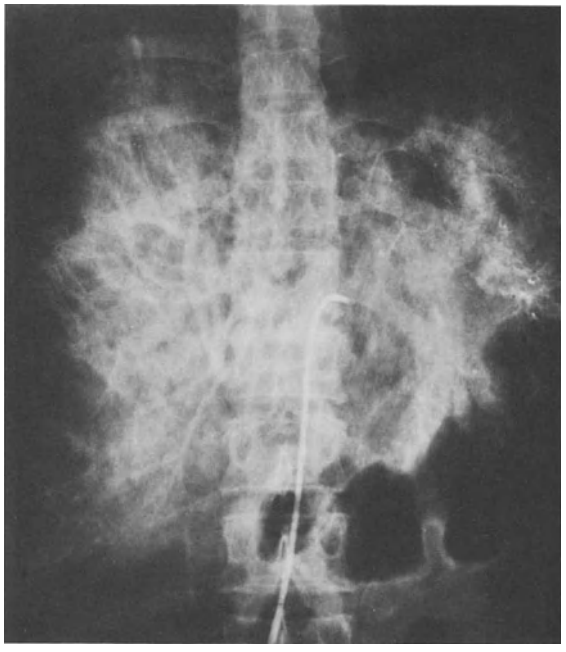


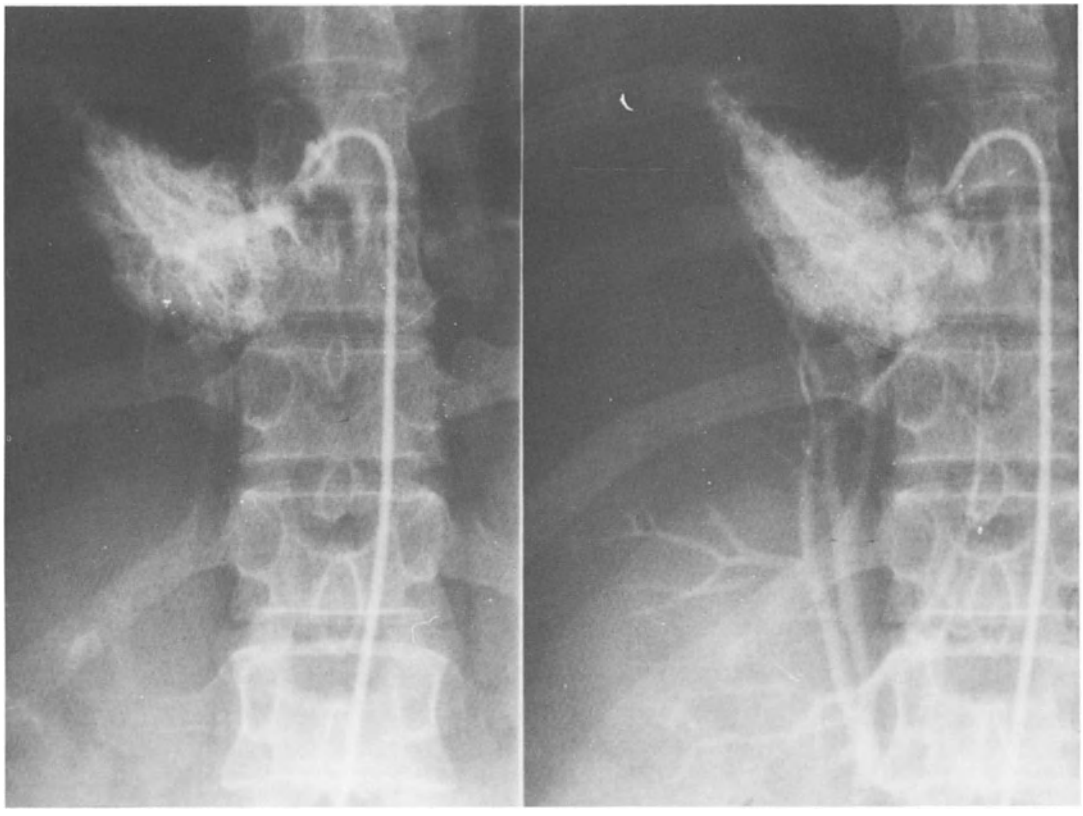
Fig. 82. Spontaneous sub-capsular hematoma of the liver revealed by abdominal pain, hepatomegaly and fever in a previously healthy 29-year-old male.

82a: Coeliac arteriography – 3rd sec – (3–10–74)
Hepatomegaly with medial displacement of the liver. Most of the right and left hepatic arterial branches are stretched and bended, suggesting a large avascular mass. Moderate hypervascular pattern in the upper right lobe where the distal arterial branches seem to be duplicated.

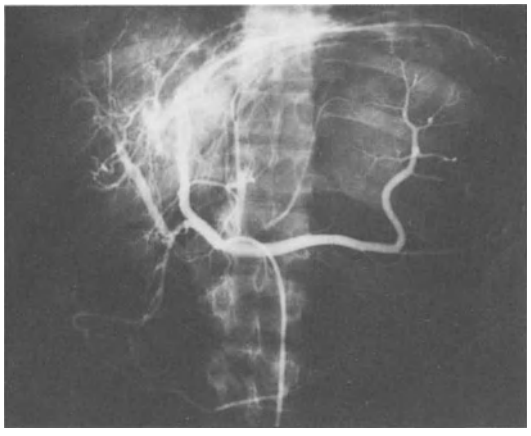
82b: Coeliac arteriography – 6th sec – (3–10–74)
Dense arterial hepatography associated with retrograde filling of the portal branches of the upper right lobe = typical pattern of the reversal of the intrahepatic portal flow in this area.

82c: Venous phase of S.M.A. – (3–10–74)
Normal portal vein with normal hepatopetal portal flow in the enlarged left lobe and the lower part of the right lobe as well. Lack of opacification of the upper right lobe where the blood flow is reversed.

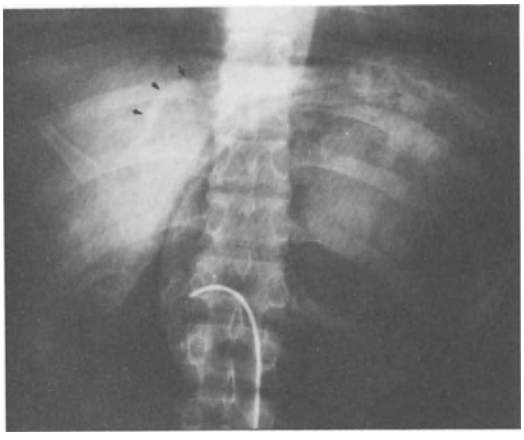
82d: Hepatic venography – (4–10–74)
The right hepatic vein is markedly narrowed and the parenchyma of the upper right lobe is greatly reduced in size due to compression by the subcapsular hematoma. Sinusoidal filling is normal but results in a retrograde opacification of the portal veins as was previously demonstrated through arterioportography.



82d



83a

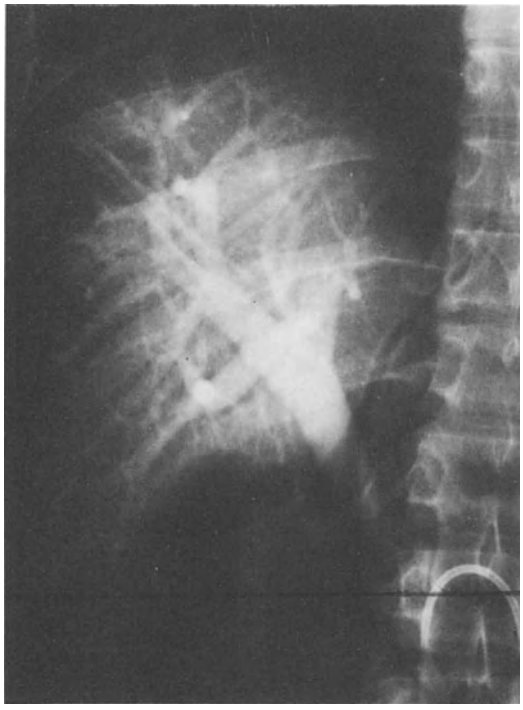


83b

Fig. 83. Pancreatitis associated with a very large pancreatic pseudo-cyst in a 20-year-old male.

83a: Coeliac arteriography – 3rd sec (16–12–75)
Upward displacement of the left branch of the hepatic artery and compression of the left lobe of the liver by the mass. The left arterial branches are slightly dilated and an abnormally dense arterial hepatography is seen in the left lobe of the liver.

83b: Coeliac arteriography – 6th sec – (16–12–75)
Retrograde filling of an intra-hepatic portal branch within the left lobe (↑). Compression of the liver parenchyma by the upper pole of the mass is well demonstrated.



84
83c

83c: Venous phase of S.M.A. (16–12–75)
Normal portal vein and right portal branch. No filling of the left portal branch where the blood flow is reversed.

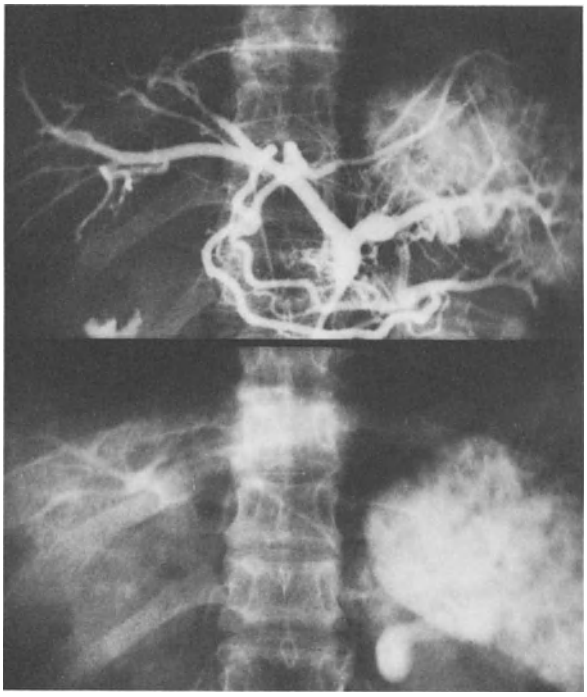
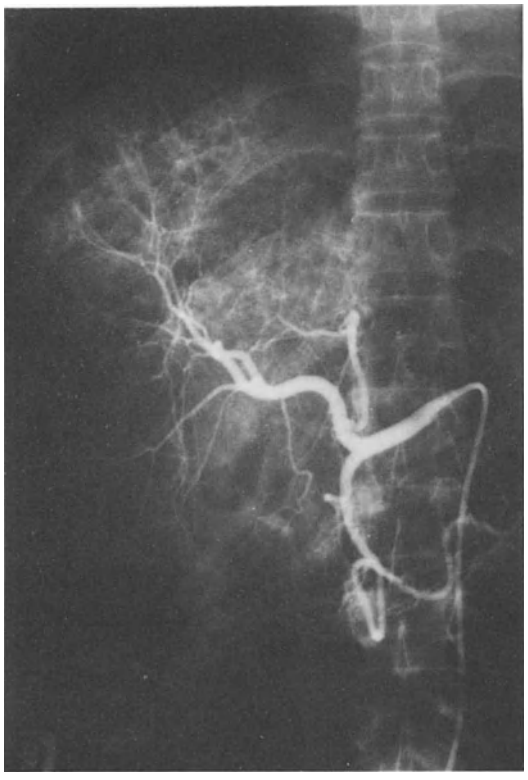


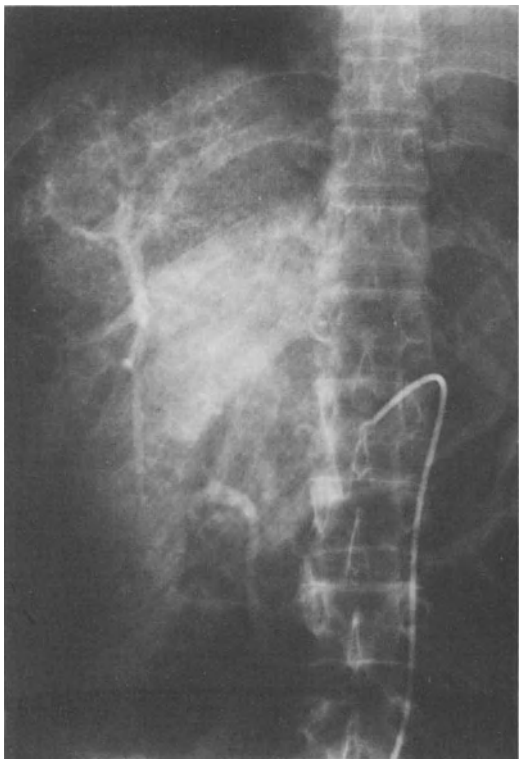
Fig. 84. Very large abdominal mass known for several years in a 28-year-old male corresponding to a voluminous retroperitoneal cystic lymphangioma. Coeliac arteriography (26–6–80) shows an upward displacement of the hepatic artery and a compression of the the left lobe of the liver (A).
6 sec later a retrograde filling of the left intrahepatic portal branches is evidenced (B). Compression of the splenic and mesenteric veins by the mass was also demonstrated.



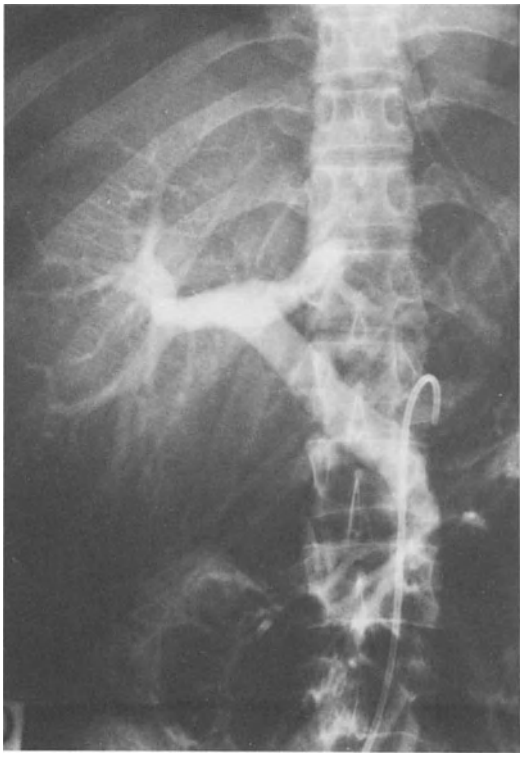
85a

Fig. 85. Hepatic trauma following a traffic accident in a 24-year-old man. Emergency surgery (13–11–75) shows a severe injury of the hepatic parenchyma of the distal part of left lobe which is treated by resection and a slight superficial injury of the upper right lobe needing only a few stitches. Arterioportography is performed 24 hours later.

85a: Selective hepatic arteriography – 3rd sec – (14–11–75)
Moderate distal hypervascular pattern in the upper right lobe (segment 7 and 8) as compared to the normal lower right lobe. A faint opacification of a few small portal venules can already be suggested in this area.

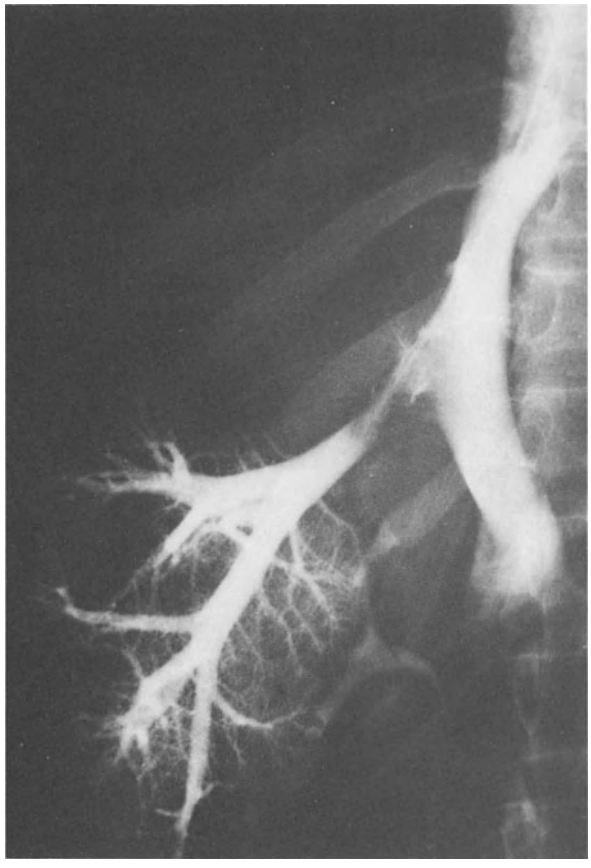


85b



85c

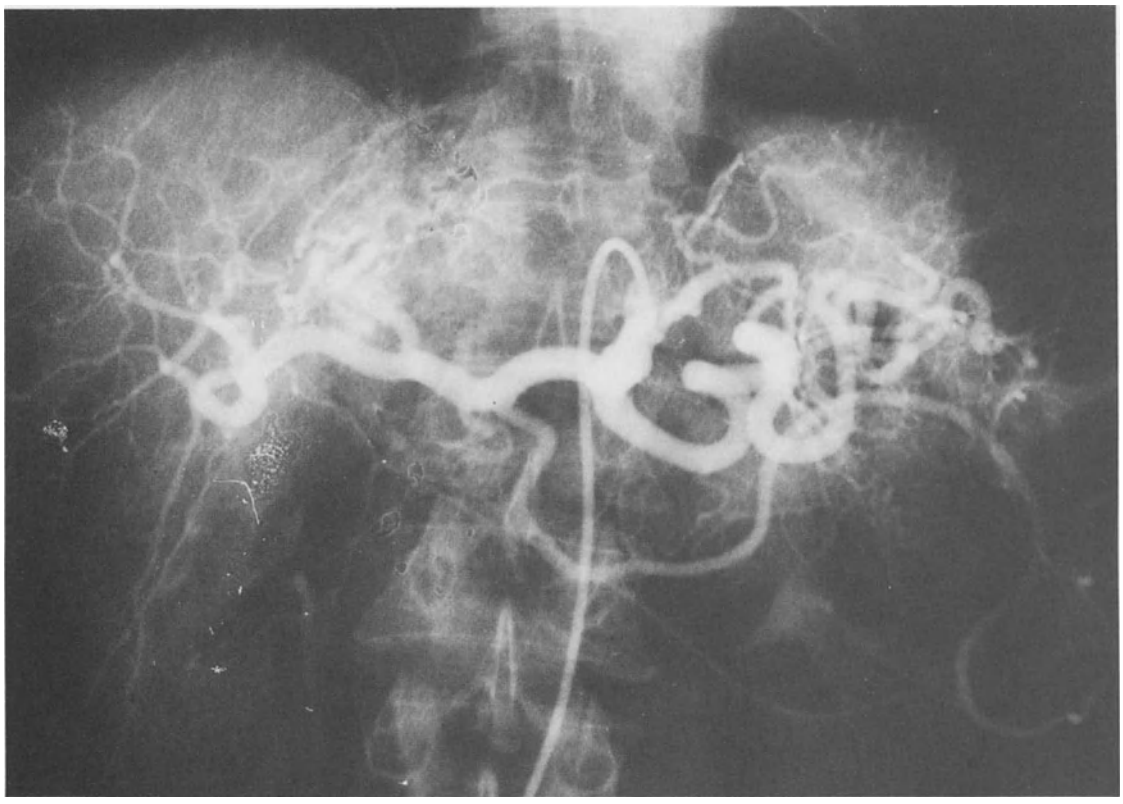
85d



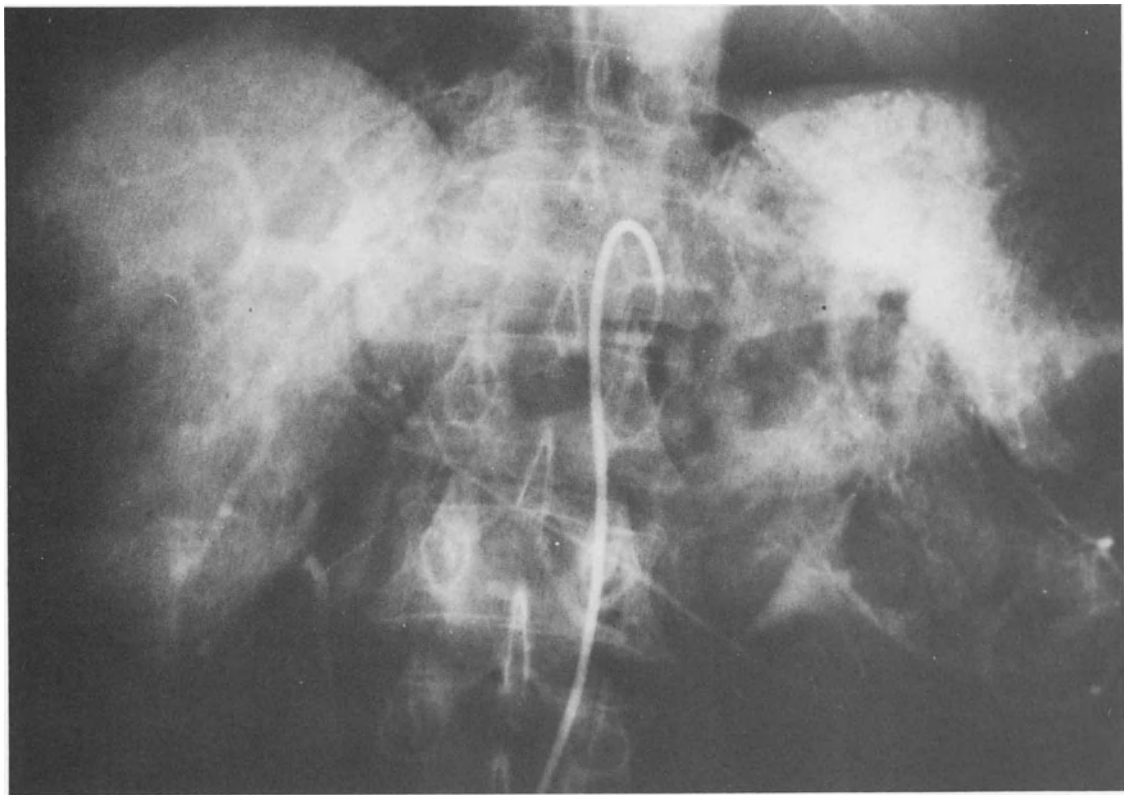
85b: Selective hepatic arteriography – 6th sec – (14–11–75)
Retrograde filling of several intrahepatic portal veins in the upper right lobe is now well evidenced.

85c: Venous phase of S.M.A. (14–11–75)
Normal portal vein with a normal hepatopetal portal flow through the lower right lobe and the remainder of the left one. Lack of opacification of the upper right lobe where the blood flow is reversed.

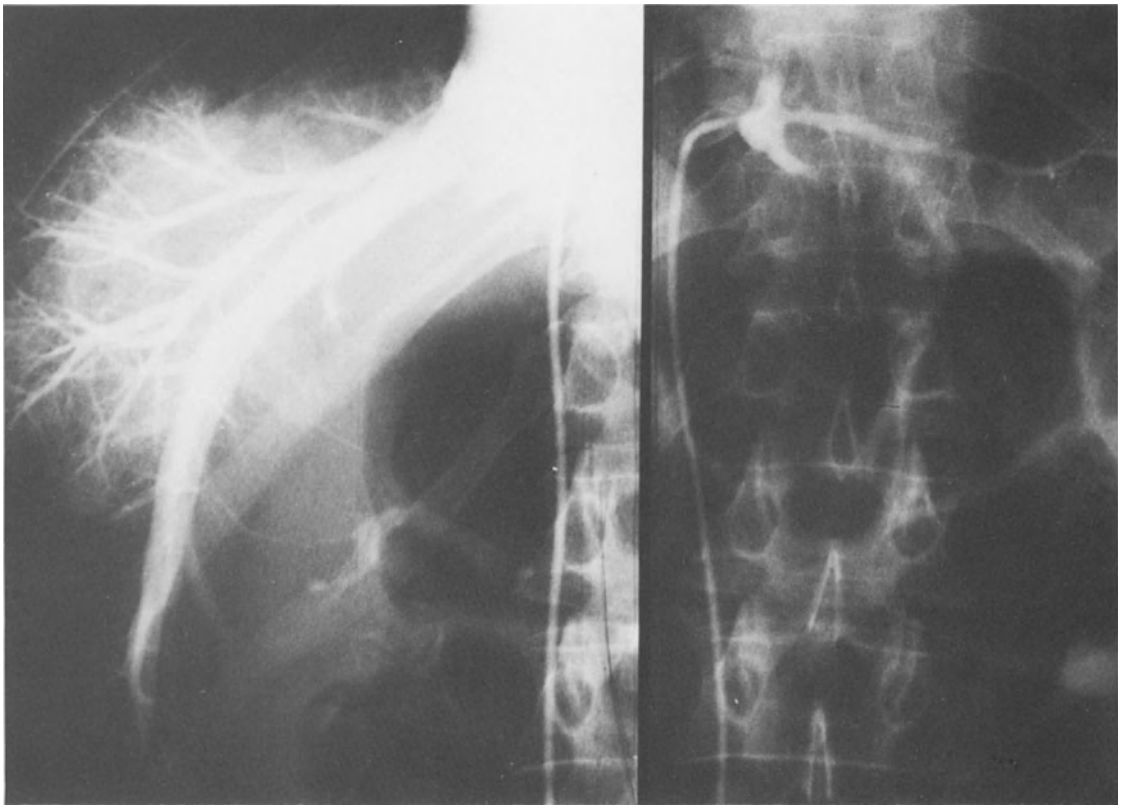
85d: Hepatic venography – (19–11–75)
Compression and downward displacement of the right hepatic vein with a normal pattern of the lower right lobe and lack of opacification of the upper right one. It was never possible to opacify the upper part of the right lobe.
Complete healing within a few days without any complication.



86a



86b



86c

Fig. 86. Chronic pancreatitis known for a few years with recurrent epigastric pain and weight loss in a 46-year-old male. Patient referred for arteriography before pancreatic surgery.

86a: Coeliac arteriography – 3rd sec – (20–10–75)

The right lobe of the liver is enlarged while the left one is small with somewhat dilated and tortuous arterial branches.

86b: Coeliac arteriography – 6th sec – (20–10–75)

Arterioportal shunting resulting in a dense retrograde opacification of the left intrahepatic portal branches.

86c: Hepatic venography – (28–10–75)

The right hepatic vein is normal whereas the left hepatic vein and its branches are markedly narrowed.

Surgical exploration discloses a hard sclerosis with atrophy of the left lobe of the liver involving the segments 1, 2 and 3 while the segment 4 (Quadratus lobe) is near normal.

Biopsy of the left lobe shows a dense sclerosis of the hepatic parenchyma with normal liver cells and a markedly thickened liver capsule.

Biopsy of the right lobe is normal.

A tumoral process can be excluded. The cause of this atrophic sclerosis of the left lobe of the liver is unknown.

GENERAL CONCLUSIONS

1) Obstruction to the portal flow is the essential phenomenon which must be evidenced through arterioportography together with its site, cause and degree, whereas portal hypertension is only an inconstant and variable consequence partly depending on the development of the collateral circulation.

2) As portal circulation is a high flow rate and low pressure gradient circulatory system, minimal intrahepatic lesions are likely to result in a mild portal hypertension which will be for a long time clinically inconspicuous until revealed by a pronounced splenomegaly.

3) In the case of an intrahepatic portal obstruction the blood flow rate is always more slowed in the splenic vein than in the superior mesenteric vein and sometimes to such an extent that the spleen cannot enlarge.

As a result, the spleen is all the more enlarged as the obstruction to the portal flow is benign and more slowly progressive whereas acute and severe obstructions are associated with a normal or near normal-sized spleen.

4) A close relationship usually exists between the decreased portal inflow and an increase of the hepatic arterial flow, except in the case of the Budd-Chiari syndrome, whose angiographic pattern compares similarly to that of an ischemic disease from venous origin. Owing to this the Budd Chiari syndrome results in a very specific angiographic pattern.

5) *Reversal of the intrahepatic portal flow* is a common finding in severe post-sinusoidal obstacles not only in cirrhosis and Budd Chiari syndrome *but in various diseases producing a focal intrahepatic obstruction whatever its cause and level.*

Arterio-portal shunting associated with the reversal of the intrahepatic portal flow should not be misinterpreted as an arterioportal fistula. They are different abnormalities which can both be evidenced through arterioportography as well as dynamic C.T.

6) When considering only the morphological changes of the liver of the spleen and of their vessels many questions about portal hypertension still remain unanswered from a physiopathological point of view. Indeed, in many cases discrepancies seem to exist between the portal pressure, the result of the liver biopsy, the morphological pattern of the liver and the spleen and the clinical background.

However, the vascular changes documented through arterioportography are likely to be closely correlated to the anatomical lesions involving the liver parenchyma.

Thus, a better understanding of the hemodynamic significance of these vascular changes should result in an improvement of our knowledge of portal flow obstruction and portal hypertension.

REFERENCES

1. Chenderovitch, J: La microangiographie hépatique l'artériographie hépatique. Premières journées Montpelliéraines de radiodiagnostic, Masson Ed, Paris, 1974.
2. Elias, H, Popper: Venous distribution in liver. Arch Path 1955, 59: 332.
3. Netter, FH: The ciba collection of medical illustrations. Vol. 3, Digestive system. Part III: Liver, biliary tract and Pancreas. 2nd edn, 1975.
4. Dock, W: The role of increased hepatic arterial flow in the portal hypertension of cirrhosis. Trans Am Physicians 1942, 57: 302–306.
5. Egbert, H, Raber, R: The physiologic effect of hepatic artery and portal ligation. Surgical Forum, Am Coll of Surgery. Philadelphia, Saunders & Cie, 1952: 153–157.
6. Mac Indoe, A: Vascular lesions of portal cirrhosis. Arch Pathol 1928, 5: 23–42.
7. Mitra, SK: The terminal distribution of the hepatic artery with special reference of arterio-portal anastomosis. J Anat 1966, 100: 651–663.
8. Geumei, AM, Mahfouz, M, Aboul-Enein, A: Intrahepatic vascular pathways. J Surg Res 1968, 8: 463–470.
9. Osteaux, M, Jeanmart, L, Drochmans, P, Bollaert, A: Conception actuelle de la microcirculation hépatique: une meilleure compréhension de l'inversion du flux portal dans la cirrhose hépatique. J Radiol Méd Nucl Electrol 1976, 57: 243–250.
10. Platteborse, R: Le foie en microscopie 'in vivo'. Librairie Maloine Editeurs, Paris, 1971.
11. Rappaport, AM: The microcirculation hepatic unit. Microvasc Res 1973, 6: 212–228.
12. Kock, NG, Hahnloser, P, Roding, B, Schenk, WG: Interaction between portal venous and hepatic arterial flow. An experimental study in the dog. Surgery 1972, 72: 414–419.
13. Lunderquist, A: Vascular anatomy of the liver. Clinical Radiology of the Liver, Part A. Herlinger, H, Lunderquist, A, Wallace, S (eds). M. Dekker, Basel, New York, 1983: 15, 53.
14. Lunderquist, A, Herlinger, H, Chuang, P: Direct portography. Clinical Radiology of the Liver, Part A. Herlinger, H, Lunderquist, A, Wallace, S (eds). M. Dekker, New York, Basel, 1983.
15. Reuter, SR, Redman, HC: Gastrointestinal angiography. Saunders Monographs in Clinical Radiology, 1972.
16. Hepatic venography and cavography. Clinical Radiology of the Liver, Part A. Herlinger, H, Lunderquist, L, Wallace, S (eds). New York, Basel, 1983.
17. Novak, D, Butzow, GH, Becker, K: Hepatic occlusion venography with a balloon catheter in portal hypertension. Radiology 1977, 122: 623–628.
18. L'Hermine, C, Paris, JC, Villoutreix, H, Dehaene, JL, Lemaitre, G: L'inversion spontanée du courant porte intra-hépatique au cours des cirrhoses. J Radiol Electrol 1975, 56: 491–503.
19. Moreno, AH, Ruzicka, FF, Rousselot, LM, Burchell, AR, Bono, RF, Slafsky, SF, Burke, JH: Functional hepatography: a study of the hemodynamics of the outflow tracts of the human liver by intraparenchymal deposition of contrast medium with attempts at functional evaluation of the outflow block concept of cirrhotic ascites and the accessory outflow role of the portal vein. Radiology 1963, 81: 65–79.
20. Mouktar, M: Hémodynamique de la circulation intrahépatique dans les cirrhoses communes: réhabilitation du rôle de l'artère hépatique dans les cirrhoses communes. Rein et Foie 1968, 11: 197–210.
21. Doyon, D, Roche, A, Chaumont, P: L'angiographie digestive chez l'adulte et chez l'enfant. Masson Ed, Paris, 1978.
22. Viamonte, M, Warren, WD, Fomon, JJ, Martinez, LO: Angiographic investigations in portal hypertension. Surg Gynecol Obstet 1970, 130: 37–53.
23. Viamonte, M, Warren, WD, Fomon, JJ: Liver panangiography in the assessment of portal hypertension in liver cirrhosis. Radiol Clin N Amer 1970, 8: 147.
24. Baum, S: Personnal Communication. 3d International Diagnostic Course, Davos, 1971.
25. Lemaitre, G, L'Hermine, C, Rembert, A, Delhay, J: Le retour veineux mésentérique supérieur au cours de l'hypertension portale. J Radiol Electrol 1972, 53 (4): 311–316.
26. Mathieu, D, Larde, D, N'Guyen, D, Bornes, JC, Vasile, N: Angioscancer hepatique normal – Méthodologie et resultats. J Radiol 1983, 64 (3): 155–160.
27. Clemett, AR, Chang, J: The radiologic diagnosis of spontaneous mesenteric venous thrombosis. Am J Gaetroenterol 1975, 63: 209–215.
28. Clarti, RA, Gallant, TE: Acute mesenteric ischemia: angiographic spectrum. AJR 1984, 142: 555–562.
29. Boijsen, E, Efsing, HO: Aneurysm of the splenic artery. Acta Radiol 1969, 8: 29–41.
30. Williams, R, Kreel, L, Blendis, L: The Liver. Butterworths, ed, 1967: 125–135.
31. L'Hermine, C, Houcke, M, Quandalle, P, Vankemmel, M: Dysplasie fibro-musculaire de l'artère splenique et hypertension portale. La nouvelle Presse Médicale, 1981, 10: 2205.
32. L'Hermine, C, Houcke, M, Quandalle, P, Vankemmel, M: Dysplasie fibro-musculaire de l'artère splenique et hypertension portale. Larc Médical, 1982, 2: 117, 123.

33. Schenk, W, Mc Donald, J, Mc Donald, K, Drapanas, T: Direct measurement of hepatic blood flow in surgical patients. Ann Surg 1962, 156: 463–471.
34. Stauffer, MH: Nephrogenic hepato-splenomegaly. Gastro-Enterology 1961, 40: 694.
35. Tartaglia, A, Wolfe, S, Prof, F, Simon: Hepatic dysfunction and fever associated with hypernephroma. NY State J Med 1970, 70: 2231–2234.
36. Niquet, G, L'Hermine, C, Dehaene, P, Defrance, G, Dupuich, Y: Syndrome de dysfonctionnement hépatique au cours d'un cancer du rein. Lille Médical, 1978, 23: 709–71.
37. Reuter, SR, Chuang, VP: The location of increased resistance to portal blood flow in obstructive jaundice. Invest Radiol 1976, 11: 54.
38. Hoevels, J, Nilsson, U: Intrahepatic vascular lesions follow non surgical percutaneous transhepatic bile duct intubations. Gastro Intest Radiol 1980, 5: 127–135.
39. Lemaitre, G, L'Hermine, C, Maillard, JP, Toison, F: L'hypertension portale segmentaire des pancréatites. Lille Médical, 1971 16: 928, 932.
40. Vachon, A, Marion, P, Jele, R, Mikaeloff, PH, de Montgolfier, R: Hypertension portale segmentaire par adénopathie bacillaire comprimant la veine splénique. Arch des Mal App Dig 1970, 59: 501.
41. L'Hermine, C, Paris, JC, Lemaitre, L, Berger, P, Delemazure, O: Complications vasculaires des volvulus chroniques du grêle. Médecine et Chirurgie Digestives 1981, 10: 113.
42. Smith-Laing, G, Camilo, ME, Dick, R, Sherlock, S: Percutaneous transhepatic portography in the assessment of portal hypertension. Gastroenterology 1980, 78: 197–205.
43. L'Hermine, C, Paris, JC, Quandalle, P, Gerard, A, Delmotte, JJ, Delhaye, JP, Paris, J: Anastomoses Porto-Caves spontanées mésenterico gonadiques au cours des cirrhoses. A propos de 3 observations. Annales de Radiologie 1972, 15: 823–829.
44. Warren, WD, Fomen, JJ, Viamonte, M, Martinez, LO, Kalser, M: Spontaneous reversal of portal veinous blood flow in cirrhosis. Surg Gynec Obstet 1968, 126: 315.
45. Kessler, RE, Tice, DA, Zimmon, DS: Retrograde flow of portal vein blood in patients with cirrhosis. Radiology 1969, 92: 1038–1042.
46. Viamonte, M, Martinez, L, Parks, RE, Warren, WD, Fomon, J: Liver shunts. Am J Radiol 1968, 102: 773–775.
47. Pollard, JJ, Nebesar, RA: Altered hemodynamics in the Budd-Chiari syndrome demonstrated by selective hepatic and selective splenic angiography. Radiology 1967, 82: 236–243.
48. L'Hermine, C, Paris, JC, Delhaye, JP, Fovet Poingt O., Lemaitre, G: L'artériographie au cours du syndrome de Budd-Chiari l'artériographie hépatique. Premières journées Montpellieraines de radiodiagnostic. Masson Ed, Paris, 1974: 147–152.
49. Galloway, S, Casarella, WJ, Price, JB: Unilobar veno-occlusive disease of the liver. Angiographic demonstration of intrahepatic competition simulating hepatoma. Am J Roentgenol 1973, 119: 89–94.
50. Maguire, R, Dopmann, JL: Angiographic abnormalities in partial Budd Chiari Syndrome. Radiology 1977, 122: 629–635.
51. Bigot, JM, Chermet, J, Monnier, JP: Les syndromes de Budd-Chiari – Diagnostic radiologique. Ann Radiol 1974, 17: 451, 469.
52. Herrick, FC: An experimental study into the cause of the increased portal pressure in portal cirrhosis. J Exp Med 1907, 9: 93–104.
53. Longmire, WP, Mulder, DG, Mahomey, PS, Mellinkoff, SW: Side-to-side porto-caval anastomosis for portal hypertension. Ann Surg 1958, 147: 881–894.
54. Leger, L, Lenriot, JP, Gorin, JP, Lemaigre, G: Néophysiologie circulatoire hépato-portale après dérivation porto-cave. J Chir 1970, 100: 457–470.
55. Reuter, SR, Orloff, MJ: Wedged hepatic venography in patients with end-to side portocaval shunts. Radiology 1974, 111: 563, 566.
56. Novak, D, Butzow, GH, Becker, K: Hepatic occlusion venography with a balloon catheter in patients with end-to-side portacaval shunts. Am J Roentgenol 1976, 127: 949–953.
57. Hoevels, J, Lunderquist, A, Tylen, U: Spontaneous intermittent reversal of blood flow in intrahepatic portal vein branches in cirrhosis of the liver. Cardiovasc Radiol 1979, 2: 267–273.
58. Kreel, L, Freston, JW, Clain, D: Vascular radiology in the Budd Chiari Syndrome. Br J Radiol 1967, 40: 755–759.
59. Cardi, J, Guille, C: Diagnostic scintigraphique et artériographique de Syndrome de Budd Chiari. Rev Méd Chir mal foie 1971, 46: 211–215.
60. Hungerford, GD, Hamlyn, AN, Lanzer, MR, Dick, R, Sherlock, S: Pseudo metastases in the liver: A presentation of the Budd Chiari Syndrome. Radiology 1976, 120: 627–628.
61. Dauver, A, Vialle, M, Coic, R, Rieux, D, Caron-Poitreau, C: Séméiologie tomodensitométrique des syndromes de Budd-Chiari. J Radiol 1982, 63: 707–716.
62. Rossi, P, Sposito, M, Simonetti, G, Sposato, S, Cusumano, G: CT diagnosis of Budd-Chiari Syndrome. J Computer Assisted Tomography 1981, 5: 366–369.
62bis. Harter, LP, Gross, BBH, St Hilaire, J, Fillg, RD, Goldblert, HI: CT and sonographic appearence of hepatic vein obstruction. Am J Roentgen, 1982, 139–176.
63. Capron, JP, Degott, C, Bernuau, J, Saint Marc Girardin, MF, Zafranie, S, Braillon, A, Quenum, C, Dhumeaux, D, Benhamou, JP: L'hyperplasie nodulaire régénératrice du foie, étude de 15 cas et revue de la littérature. Gastroenterol Clin Biol 1983, 7: 761–769.
64. Levin, DM et al.: La fréquence des hépatopathies non alcooliques est sous estimée chez les éthyliques. Am J Med 1979, 66: 429–434.
65. L'Hermine, C, Paris, JC, Dehaene, JL, Rondot, P: Les cirrhoses non éthyliques. Apport de l'arteriographie sur le plan diagnostique et éthiopathogénique. A propos de 80 observations. Ann Radiol 1975, 18: 679–688.
66. Buligesco, L, Paunesco, C: L'hypersplénisme, séquelle

d'hepatite épidémique. Acta Gastro-Enterologica Belgica 1966, 29: 791.
67. Gherman, Gr, Papilian, VV: Quelques aspects de la splénopathie dans l'hépatite chronique. Revue Internationale d'Hépatologie 1965, 15: 1215.
68. Fodor, O, Stanesco, L, Cosma, V, Itu, I, Fratila, I, Munteanu, P, Schwartz, M, Ciofu, Gh, Ilea, V, Cotul, S, Dumitrasco, D, Borsan, I: Investigations cliniques et épidémiologiques sur le passage à la chronicité de l'hepatite épidémique aiguë. Revue Internationale d'Hépatologie 1964, 14 (6), 567.
69. Kunkel, HG, Labby, DH: Chronic liver disease following infectisis hepatitis. II – Cirrhosis of the liver. Ann Int Med 1950: 32, 433.
70. Davis, WD Jr: The sequelae of hepatitis. Am J Dig Dis 1964, 9: 811.
71. Chiandussi, L, Greco, F, Indovina, D, Cesano, L, Vaccarino, A, Muratori, F: Hepatic stéatosis and portal hypertension with presinusoidal obstruction. Report of a case. Gastroenterology 1963, 44: 532.
72. Bourel, M, Simon, M: Le point sur l'hémochromatose idiopathique. Arch Fr Mal App Dig 1975, 64: 385.
73. Darnis, F: Les gros foies de Surcharge. Revue du Praticien 1963, 13: 77.
74. Farmer, RG, Sullivan, BH: Portal hypertension in hémochromatosis. Am J Dig Dis 1964, 9: 56.
75. Leger, L, Lemaigre, G, Premont, M, Salmon, R, Klioua, Z, Battesti, JP: Hypertension portale au cours de la sarcoïdose. Nouvelle Presse Méd 1980, 9: 1021.
76. Koppich, Fr, Georgesco, I St, Blanc, G, Dimitriu, M: Considérations préliminaires sur l'évolution de l'hépatite chronique postvirale. Acta Gastro-Enterologica Belgica 1963, 26: 854.
77. Runcan, V, Vulpesco, S, Marcus, N, Popesco, T: Les perturbations de l'hémodynamique portale au cours des hépatites chroniques consécutives à l'hépatite épidémique. Revue Internationale d'Hépatologie 1966, 16: 997.
78. Lebrec, D, Nouel, O, Bernuau, J, Benhamou, JP: L'hépatite virale fulminante est toujours associée à une hypertension portale. Gastroentérol Clin Biol 1980, 4(Ibis): 107A.
79. Sherlock, S, Feldman, CA, Moran, B, Schever, PJ: Partial nodular transformation of the liver with portal hypertension. Am J Med 1964, 40: 195.
80. Shedlofsky, S, Koehler, RE, Deschryver, Kecskemeti, K, Alpers, DH: Non cirrhotic nodular transformation of the liver with portal hypertension. Clinical, angiographic and pathological correlations. Gastro-enterology 1980, 79: 938, 943.
81. Futagawa, S, Fukazawa, M, Horisawa, M, Muska, H, Ito, T, Suziura, M, Kameda, H, Okuda, K: Portographic liver changes in idiopathic noncirrhotic portal hypertension. Am J Roentgenol 1980, 134: 917–923.
82. Mikkelson, WP, Edmondson, HD, Peters, RI, Redecker, A, Reynolds, TB: Extra and intra-hepatic portal hypertension without cirrhosis (hepatoportal scleroses). Ann Song 1963, 162: 602, 620.
83. Leger, L, Richarme, J, Lemaigre, G: L'hypertension portale essentielle. Archives des Mal App Dig 1965, 54: 153.
84. Kreis, H: Les complications hépatiques de la transplantation rénale. Annales de gastroentérologie et d'hépatologie 1979, 15: 337–340.
85. Nataf, C, Feldmann, G, Lebrec, D, Degott, C, Descamps, JM, Rueff, B, Benhamou, JP: Idiopathic portal hypertension (perisinusoidal fibrosis) after renal transplantation. Gut 1979.
86. Varay, A: Que reste-t-il des cirrhoses d'origine splénique? Revue Internationale d'Hépatologie 1962, 12: 34.
87. Bourgeon, R, Alexandre, JH, Mouiel, J: La splénomégalie au cours des cirrhoses méditerranéennes. Conception d'ensemble. Déductions thérapeutiques. Arch Fr Mal App Dig 1966, 55: 507.
88. Kameda, H, Takeda, J: Patho physiology of idiopathic portal hypertension. J Ap J Med 1969, 8: 29–31.
89. Boyer, JE, Hales, MR, Klatskin, G: Idiopathie portal hypertension due to occlusion of intrahepatic portal veins by organized thrombi. Medicine 1974, 53, 77.
90. Villeneuve, JP, Huet, PM, Joly, JC, Marleau, D, Cote, J, Legare, A, Lafortune, M, Lavoie, P, Viallet, A: Idiopathic portal hypertension. Am J Med 1976, 61: 459–464.
91. Payet, M, Menache, D, Saimot, G, Schlegel, N, Eyquem, P, Coulaud, JP: L'antigène Australie chez l'Africain. Presse Médicale 1971, 79: 2359.
92. Labayle, D, Lenoir, C, Buffet, C, Chaput JC, Etienne, P: Hépatites actives d'origine médicamenteuse. Nouv Presse Méd 1979, 8: 2013–2015.
93. Nalpas, B, Berthelot, P: Complications hépatiques des traitements médicamenteux. Concours Médical 1981, 103: 1663–1673.
94. Poupon, R, Longchal, C, Darnis, F: Hépatite chronique associée à la prise prolongée de papavérine. Gastroentérol Clin Biol 1978, 2: 305–308.
95. Tassou, JJ, Bretagne, JF, Marion, J: Hépatite à la clométacine. Concours Médical 1981, 103: 679–683.
96. Leger, L, Mouktar, M, Alfandari, JP: Hypertension portale d'amont, d'apport ou de surcharge. Presse Médicale 1965, 73: 479.
97. Viamonte, M, Damer, P, Warren, WD, Fomon, J: A new technique for the assessment of hypertinetic portal hypertension. Radiology 1970, 96: 539.
98. Chuang, VP, Lunderquist, A, Herlinger, H: Portal hypertension. Clinical radiology of the liver. Part B. Marcel Dekker, New York, Basel, 1983.
99. Van Way, CW, Crani, JW, Riddell, DH, Foster, JH: Arteriovenous fistula in the portal circulation. Surgery 1971, 70: 867–90.
100. Maillard, JN, Benhamou, JP, Rueff, R: Les fistules arterio-portales. Aspects cliniques et physiopathologiques à propos d'un cas. Arch Fse des Mal App Disgestif 1968, 57: 1017, 1032.
101. Puglionisi, A, Digiovani, V, Snider, F, Camills Cina, GA: A propos d'un cas de fistule artério-veineux hépaticoportale – Problèmes de Physiopathologie et clinique et revue de la littérature. J Chir 1980, 117: 607–619.
102. Schilling, JA, McKee, FW, Wilt, W: Experimental hepatico-portal arterio-venous anastomosis. Surg Gynecol

Obstet 1950, 90: 473, 479.
103. Morand, P, Potier, N, Barsotti, J, Raynaud, R: Hypertension portale par anevrysme artérioveineux des vaisseaux gastro épiploïques droits après gastrectomie. Sem Hop Paris 1973, 49: 1837, 1843.
104. Schwartz, SI, Martin, JH, Mac Govern, GR: Experimental arterialisation of the liver. Surgery 1961, 49: 611.
105. Mallet, WYP, Kayababi, I, Fesoldi, J, Mally, GY: Artérialisation expérimentale de la veine porte. Données histologiques et ultrastructurales. Presse Médicale 1968, 76: 1221, 1225.
106. Madding, GF, Smish, NL, Herberger, LR: Hepatoportal arteriovenous fistula. JAMA 1954, 156–593.
107. Shumaker, HB, Waldhauser, JA: Intra hepatic arterioveinous fistula of the hepatic artery and portalvein. Surg Gynec Obstet 1981, 112–497.
108. Wheeler, HB, Warren, R: Duodenal varices due to portal hypertension from arteriovenous aneurysm. Ann Surg 1957, 146–229.
109. Capron, JP, Gineston, JL, Remond, A, Lallement, PY, Delamarre, J, Revert, R, Veyssier, P: Inferior mesenteric arterioveinous fistula associated with portal hypertension and acute ischemic colitis – successful occlusion by intra arterial embolization with steel coils. Gastro enterology 1984, 86: 351, 355.
110. Adler, J, Goodgold, M, Mitty, H, Gordon, D, Kinkhabwala, M: Arteriovenous shunts involving the liver. Radiology 1978, 129: 315–322.
111. L'Hermine, C, Vankemmel, M, Quandalle, P, Dehaene, JL, Maillard, JP: Le contrôle de la perméabilité des anastomoses portocaves chirurgicales par l'artérioportographie. J Radiol Electrol 1976, 57: 219–225.
112. Reuter, SR, Berk, BN, Orloff, MJ: An angiographic study of the pre and post-operative hemodynamics in patients with side-to-side portocaval shunts. Radiology 1975, 116: 33–39.
113. Drapanas, TH: Interposition mesocaval shunt for treatment of portal hypertension. Ann Surg 1972, 176: 435.
114. Drapanas, TH, Locicero, J, Dowling, JB: Hemodynamics of the interposition mesocaval shunt. Ann Surg 1975, 181: 523.
115. Fulenwider, JT, Nordlinger, BM, Millekan, WJ, Jones, WJ, Warren, PJ: Portal pseudoperfusion – An angiographic illusion. Ann Surg 1979, 189: 257.
116. Herlinger, H, Chuang, P, Lunderquist, A: Arterioportography. Clinical Radiology of the Liver. Part A. Herlinger, H, Lunderquist, A, Wallace, S (eds). New York, Basel 1983: 337.
117. Ribet, M, Paris, JC, Quandalle, P, Chambon, JP, Houcke, P: Place de l'anastomose mésentérico-cave dans le traitement de l'hémorragie par hypertension porte. Méd Chir Dig 1981, 10: 121–126.
118. Witte, CL, Ovitt, TW, Witte, ML, Clark, DS: Left sided segmental portal hypertension following mesocaval interposition shunt. Surg Gynecol Obstet 1977, 145: 169–174.
119. Warren, WD, Millikan, WJ, Henderson, JM et al.: Ten years portal hypertensive surgery at emory – results and new perspectives. Ann Surg 1982, 195: 530–542.
120. Maillard, TN, Flamant, YM, Hay, JM, Chandler, JG: Selectivity of the distal spleno-renal shunt. Surgery 1979, 86: 663.
121. Tylen, V, Simert, G, Vang, J: Hemodynamic change after distal splenorenal shunt studied by sequential angiography. Radiology 1976, 121: 585.
122. Widrich, WC, Robbins, AH, Nalseth, DC, O'Hara, ET, Johnson, WC, Longhlim, KV: Portal hypertension changes following selective spleno-renal shunt surgery. Radiology 1976, 121: 295–302.
123. Benacerraf, R, Vermelin, PH, Daniel, B: L'anastomose spléno-rénale distale de Warren. J Radiol 1979, 60: 493–502.
124. Nordlinger, BM, Nordlinger, DF, Fulenwider, JT et al.: Angiography in portal hypertension clinical significance in surgery. Am J Surg 1980, 139: 132–141.
125. Warren, WD, Restrepo, JE, Respess, JC, Muller, W: The importance of hemodynamic studies in management of portal hypertension. Ann Surg 1963, 158: 387.
126. Smith, GW: Use of hemodynamic selection criteria in the management of cirrhotic patients with portal hypertension. Ann Surg 1974, 179: 782.
127. Imanaga, H, Yamamoto, S, Kuroyanagi, Y: Surgical treatment of portal hypertension according to state of intrahepatic circulation. Ann Surg 1962, 155: 42–50.
128. Hammers, LW, Glanz, S, Cordon, DH, Sclafani, JA, Adamsons, RJ: Arterialization of the portal vein for control of variceal bleeding. Radiology 1983, 149: 649–653.
129. Maillard, JN, Rueff, B, Prandi, D, Sicot, C: Hepatic arterialization and portacaval shunt in hepatic cirrhosis. An assessment. Arch Surg 1974, 108: 315–320.
130. Chastanet, P: Embolisation des varices oesophagiennes par voie transhépatique percutanée. Bilan de notre expérience chez 300 malades. Thèse, Lille, 1983.
131. Bengmark, S: The surgeon's viewpoint. Clinical Radiology of the Liver. Part B. Herlinger, H, Lunderquist, A, Wallace, S, Dekker, M (eds). New York, Basel 1983: 595.
132. Maillard, JN, Flamant, Y: L'hypertension portale du cirrhotique justifie-t-elle encore une anastomose porto-cave? Bordeaux Medical 1979, 12: 1643.
133. Vankemmel, M, Paris, JC, Lagache, G: Décompression portale hyper-sélective pour rupture de varices oeso-cardiales. Chirurgie 1982, 108: 657–666.
134. Vankemmel, M, Francke-Mauroy, B, Aubineau, JM: La résection-anastomose de l'oesophage sus-cardial pour rupture de varices oesophagiennes. Méd Chir Dig 1982, 11: 111–115.
135. Lunderquist, A, Vang, J: Transhepatic catheterisation and obliteration of the coronary vein in patients with portal hypertension and oesophageal varices. N Engl J Med 1974, 291: 646–659.
136. Sos, TA: Transhepatic portal venous embolization of varices: Pros and cons. Radiology 1983, 148: 569–570.
137. Widrich, WC, Robbins, AH, Nabseth, DC, Johnson, WC, Golstein, SA: Pitfalls of transhepatic portal venography and therapeutic coronary vein occlusion. Am J Roentgenol 1978, 131: 637–643.
138. Levade, M, Vinel, JP, Railhac, JJ, Paille, P, Pascal, JP,

Putois, J: Embolisation au bucrylat des varices oeso-cardio-tuberositaires par voie transjugulaire chez le cirrhotique. 5e Congres Europeen de Radiologie, Bordeaux 5–10 Septembre 1983.

139. Lunderquist, A, Simert, G, Tylen, U, Vang, J: Followup of patients with portal hypertension and esophageal varices treated with percutaneous obliteration of gastric coronary vein. Radiology 1977, 122: 59–63.
140. L'Hermine, C, Chastanet, P: Embolisation des varices oesophagiennes par voie transhepatique percutanee. Résultats chez 300 patients. Ann Radiol 1984, 27: 292–293.
141. Mc Fadzean, AJ, Cook, J: Ligation of the splenic and hepatic arteries in portal hypertension. Lancet 1953, 1: 615–622.
142. Williams, R, Condon, RE, Williams, HJ et al.: Splenic blood flow in cirrhosis and portal hypertension. Clin Sci 1968, 34: 441–452.
143. Bücheler, E, Thelen, M, Schirmer, G et al.: Katheterembolisation du Milzarterien zum stopp der akuten varizenblutung. Fortsch – Geb Roentgenstr Nuklearmed 1975, 122: 224–229.
144. Dumont, AE, Berman, IR, Stohl, NM et al.: Significance of an enlarged splenic artery in patients with bleeding varices. Ann Surg 1972, 175: 466–471.
145. Witte, CL, Witte, MH, Ressert, W. et al.: Splenic artery ligation in selected patients with hepatic Cirrhosis and in sprague-Dawley rats. Surg Gynecol Obstet 1976, 142: 1–12.
146. Owman, J, Lunderquist, A, Alwmark, A, Borjesson, B: Embolization of the spleen: treatment for hypersplenism in the patient with portal hypertension. Presented at the World Congress of Gastro enterology – Madrid, June 1978.
147. Gerlock Jr, AJ, MacDonell Jr, RC, Muhletaler, CA, Parris, WCV, Johnson, HK, Tallent, MB, Richie, RE, Kendall, RI: Partial spenic embolization for hypersplenism in renal transplantation. Am J Roentgenol 1982, 138: 451, 456.
148. Goldman, ML, Philip, PK, Sarrafizadeh, MS, Sarfeh, J, Salam, AA, Galambos, JT, Powers, SR, Balint, JA: Intra-arterial tissue adhesive for medical splenectomy in humans. Radiology 1981, 140: 341–349.
149. Castaneda Zuniga, WR, Hammerschmidt, E, Sanchez, R, Amplatz, K: Nonsurgical splenectomy. Am J Roentgenol 1977, 129: 805–811.
150. Anderson, JH, Vuban, A, Wallace, S, Hester, JP, Burke, JS: Transcatheter splenic arterial occlusion: an experimental study in dogs. Radiology 1977, 125: 95–102.
151. Porter, BA, Frey, CF, Link, DP, Lantz, MT, Pimstone, NR: Splénic embolization monitored by the video dilution technique. Am J Roentgenol 1983, 141: 1063–1065.
152. Conn, HO, Ramsby, GR, Storer, EH, Mutchnick, ML, Joshi, PH, Philips, MM, Cohen, GA, Fields, GN, Petroski, D: Intra arterial vasopressin in the treatment of upper gastrointestinal hemorrhage: a prospective controlled clinical trial. Gastro enterology 1975, 68–211.
153. Sivak Jr, MV: Endoscopic injection sclerosis of esophageal varices: A/S/G/E Survey (letter) Gastrointest Endosc 1982, 28: 41–42.
154. Johnson, GW, Rodgers, HW: A review of 15 years' experience in the use of sclerotherapy in the control of acute haemorrhage from esophageal varices. Sclerotherapy in acute haemorrhage. Br J Surg 1973, 60: 797–799.
155. Colapinto, RF, Stronell, RD, Gildiner, M, Ritchie, AC, Langer, B, Taylor, BR, Blendis, LM: Formation of intrahepatic porto systemic shunts using a balloon dilatation catheter: preliminary clinical expericence. Am J Roentgenol 1983, 140: 709–714.
156. Axel, L, Moss, AA, Berninger, W: Dynamic computed tomography demonstration of hepatic arterio-venous fistula – case report. J Comput Assist Tomogr 1981, 5: 95–98.
157. Inamoto, K, Tanaka, S, Yamazaki, H, Hayashi, T, Hikada, H, Miura, K: Arterioportal fistula in hepatocellular carcinoma. J Comput Assist Tomogr 1983, 7: 151, 153.
158. Mathieu, D, Larde, D, Vasile, N: CT Features of iatrogenic hepatic arterioportal fistulae. J Comput Assist Tomogr 1983, 7: 810–814.
159. Baer, JW: Hepatic arterioportal fistula related to a liver biopsy. Gastrointest Radiol 1977, 2: 297–299.
160. Nakayama, Hiyama, Y, Ohnishi, K, Tsuchiya, S, Kohno, K, Nakajima, Y, Okuda, K: Arterioportal shunts on dynamic computed tomography. Am J Roentgenol 1983, 140: 953–957.
161. Hoiem, L, Kvam, G: Arterio-portal fistula diagnosed by computerized tomography (CT). Eur J Radiol 1981, 1: 57–59.
162. Bookstein, J, Boijsen, E, Olin, T, Vang, J: Angiography after end-to-side porto-caval shunt (clinical, laboratory and pharmaco-angiographic observations). Invest Radiol 1971, 6: 101.
163. Mac Loughlin, MJ, Quinn Hill, M: Angiography in cholangio carcinoma complication hemochromatosis. J Ass Canad Radiol 1970, 21: 238.
164. Sniderman, KW, Sos, TA: Hepatic schistosomiasis: A case with intra hepatic shunting and extra hepatic portal vein occlusion. Am J Roentgen 1978, 130: 565.
165. Paris, JC, L'Hermine, C, Tonnel, AB, Dehaene, JL, Plouvier, B, Pagniez, B, Pommelet, P: Fistule artério-porte-hépatique secondaire à une thrombose porte au cours d'une cirrhose. J Radiol Electrol 1976, 4: 227–232.
166. Delemazure, O: Etude angiographique des communications artério-portes intra-hépatiques (à propos de 66 observations). Thèse pour le Doctorat en Médecine, Université de Lille, 1979.
167. L'Herminé, C, Paris, JC, Dehaene, JL, Villoutreix, H, Denies, JL: Inversion du courant portal par obstruction des veines sus-hépatiques au cours des tumeurs du foie. J Radiol Electrol 1975, 56: 299–306.
168. Moss, AA, Dean, PB, Axel, L, Goldlez, HI, Glazer, G, Friedman, MA: Dynamic C.T. of hepatic masses with intravenous and intraarterial contrast material. AJR, 1982, 138: 847–852.
169. Itai, Y, Moss, AA, Goldberg, HI: *Transient* hepatic attenuation difference of lobar or segmental distribution detected by dynamic computed tomography. Radiology 1982, 144: 835.

170. L'Herminé, C, Vankemmel, M, Dehaene, JL, Houcke, M, Viaud, A: Hématome sous-capsulaire spontané du foie comprimant les veines sus-hépatiques. Diagnostic angiographique. Journal de Radiologie 1976, 57: 747–752.
171. L'Herminé, C, Grand, M, Remond, A, Vankemmel, M, Quandalle, P: L'inversion du courant porte intra-hépatique au cours des contusions du foie: une nouvelle conception des 'fistules artério-portes' traumatiques. Journal de Radiologie 1977, 58: 609–619.
172. Boijsen, E, Kaude, J, Tylen, U: Angiography in hepatic rupture. Acta Radiol 1971, 11: 363–379.
173. Okuda, K, Suzuki, K, Musha, H, Arimizu, H: Percutaneous transhepatic catheterization of the portal vein for the study of portal hemodynamics and shunts. A preliminary report. Gastroenterology 1977, 73: 279–284.