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Volume 19

Long-gap Esophageal Atresia

Prenatal Diagnosis of Congenital Malformations

Volume Editor
P. Wurnig, Vienna

With 86 Figures

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In Memoriam Professor Francesco Soave

Here we would like to turn our thoughts to Francesco Soave and to express our deep gratitude to him.

Francesco Soave was born on June 14, 1917, in Meta di Sorrento and died in Genoa on September 26, 1984. He was appointed surgeon in chief at the I. G. Gaslini Institute Hospital in 1955 and professor of pediatric surgery at the University of Genoa in 1966, holding both positions until his death. His special field was colonic and anorectal surgery, and he often gave demonstrations and lectures on it in many countries as a visiting or guest professor.

Professor Soave became a member of the British Association of Paediatric Surgery in 1957 and organized its annual congress in Genoa in 1971. He was the Windermere Foundation Travelling Professor of Surgery in Australia in 1978 and the overseas guest speaker at the annual meeting of both the surgical section of the American Association of Pediatrics (1971) and the American Pediatric Surgical Association (1984). He held the Forshall Lecture in 1971 and the Oberniedermayr Lecture in 1975 as a member of the Deutsche Gesellschaft für Kinderchirurgie.

Professor Soave served on the editorial boards of Progress in Pediatric Surgery, Zeitschrift für Kinderchirurgie, and the Journal of Pediatric Surgery. He was president of the Societa Italiana di Pediatria from 1970 to 1972.

Innumerable international honorary presentations, awards, and honorary memberships were devoted to him by pediatric and pediatric surgical societies. This outstanding pediatric surgeon, teacher, and scientist wrote more than 160 publications.

The pediatric surgery family has lost not only one of its most eminent members, but also a deep and cordial friend.

THOMAS A. ANGERPOINTNER/WALDEMAR CH. HECKER, Munich

Preface

The comparison of established methods in surgery is necessary in order to evaluate the advantages or disadvantages of each. We have therefore tried to include discussions of all the problems which arise in the treatment of long-gap esophageal atresia.

The long-term results of different types of colonic interposition, of different "stretching" procedures, and of simple staged surgery seemed especially worthy of discussion. It was also important to describe the role of complications caused by special pathology of the trachea in esophageal atresia and their management.

Second, new problems continue to arise with regard to the prenatal diagnosis of malformations. These new aspects will continue to exert an influence on our surgical field. Malformations pose severe problems for parents, the growing fetus, and the doctors and are lasting burdens on our task.

PETER WURNIG, Vienna

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You will find the adresses at the beginning of the respective contribution

Current Surgical Strategies in Long-gap Esophageal Atresia with Regard to Endoscopic Anastomosis

D. Booss[1] and J. Kotlarski[1]

Pediatric surgeons have long been engaged with the problem of long-gap esophageal atresia. The methods of treatment can be roughly subdivided into (a) interposition and (b) elongation.

Interposition methods are well known and widely in use. Colonic interposition according to Waterston (1964) is appreciated and employed. After 20 years we have enough experience to make a critical evaluation of the method; some questions remain as far as long-term results are concerned. It was therefore necessary to develop methods which avoid the interposition of material originating from another region of the body.

Elongation methods have succeeded more and more during the past 10 years. They are based on the principle of elongating the pouches to such an extent that connection is possible. These methods are linked with the names Howard and Myers, Rehbein, Hendren, Livaditis, and Hecker (Howard and Myers 1965; Rehbein and Schweder 1971; Rehbein 1976; Hendren and Hale 1975; Livaditis et al. 1972; Daum et al. 1970).

We report on seven cases of long-gap esophageal atresia from a total of 50 children who were operated on for esophageal atresia at the Pediatric Surgical Clinic of Bremen from 1976 to 1983 (see Table 1). Four children presented with type IIIa and three with type II, according to Vogt's classification. In one of the children with type IIIa esophageal atresia and a surgically closed upper fistula, bougienage according to Howard and Myers (1965) could not be started for 3 months due to initial cardiopulmonary complications connected with Down's syndrome, duodenal stenosis, and severe congenital heart disease. Thereafter, we proceeded with bougienage, and anastomosis could be performed without difficulties. The esophageal passage was free and oral feeding was started with caution. The child died of pulmonary and septic complications, however, 2 months postoperatively.

The remaining children underwent elongation treatment according to Howard and Myers (1965) for 5 weeks to 3 months; in one child (patient no. 5) only the lower pouch could be elongated owing to a complication following closure of the upper fistula. In the remaining infants both pouches were elongated (Fig. 1). The table shows the length of the gap in either cm or measures of vertebral bodies. The gaps were between two and four vertebral bodies, or roughly 2 cm.

In five cases we performed primary anastomosis of the pouches after roentgenograms had revealed sufficient closing over a period of several weeks or months (Fig. 2).

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Table 1. Long-gap esophageal atresia cases treated at the Pediatric Surgical Clinic, Bremen 1976–1983

No.	Patient's birth weight	Vogt's classification	Length	Therapy					Complications			
				Howard and Myers	Thread	Olives	Anastomosis		Stenosis	Others	Result	Remarks
							Primary	Secondary				
1	BP, ♂, 2960 g	IIIa	3½ vb	+	+ endoscopically	+	–	+	+	–	Good	
2	DS, ♂, 2000 g	II (IIIa)	2 cm	+	+ endoscopically	+	–	+	+	–	Good	Gastroesophageal reflux
3	RS, ♀, 1600 g	II	2.5 cm	+	–	–	+	–	(+)	–	Good	
4	RD, ♂, 1570 g	II	2 cm	+	–	–	+	–	(+)	–	Good	Trisomy 21, gastroesophageal reflux 3½ years later
5	KB, ♂, 1510 g	IIIa	2 vb	+	–	–	+	–	–	–	Good	
6	BP, ♂, 3050 g	IIIa	2½ vb	+	–	–	+	–	–	–	Good	
7	LL, ♀, 2230 g	IIIa	4 vb	+	–	–	+	–	–	–	Died	Trisomy 21, vitium cordis, duodenal stenosis

vb = Vertebral bodies

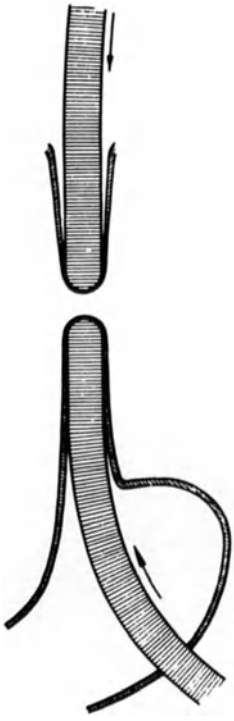
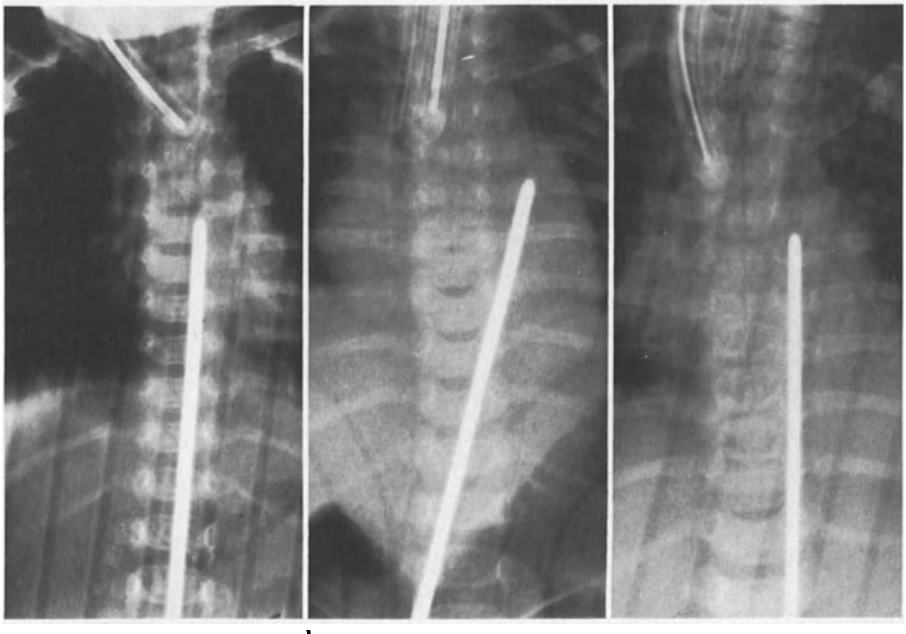


Fig. 1. Bougienage according to Howard and Myers



a **b** **c**

Fig. 2a–c. Stages of approach of the esophageal pouches (patient no. 6) **a** after 4 weeks, **b** after 8 weeks, **c** after 12 weeks

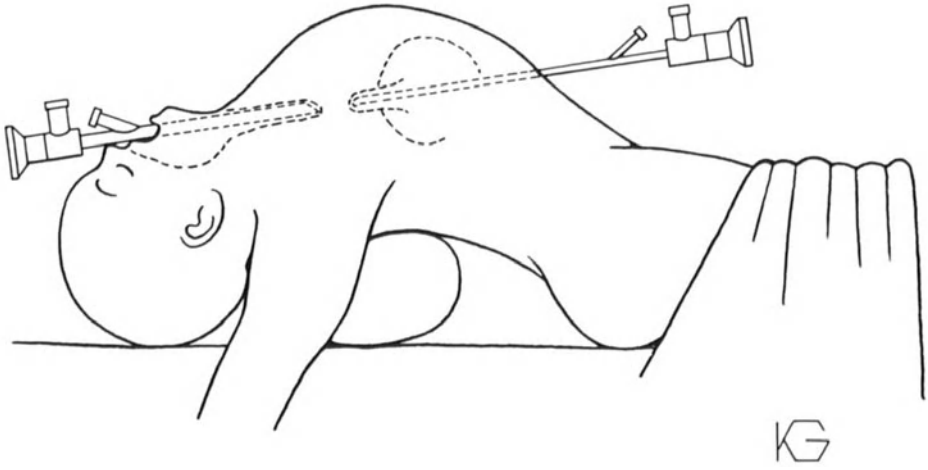


Fig. 3. Position of patient and of the two endoscopes for inserting the thread

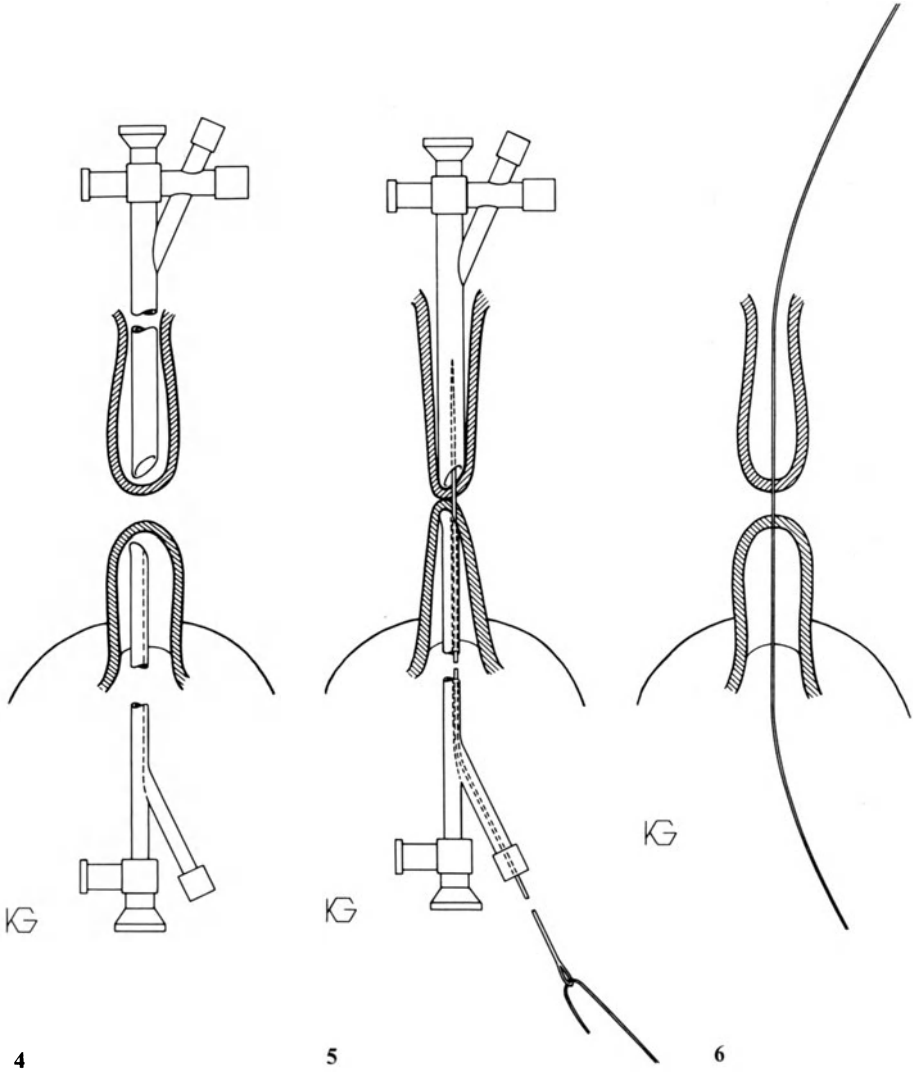
In another two cases we employed elongation by means of the olive-and-thread method prior to anastomosis; the nylon thread was inserted endoscopically. Via an endoscope inserted in the gastrostoma a long sterile needle with the thread attached was advanced from the lower to the upper pouch, where it entered a stiff esophagoscope and was led to the exterior through the mouth. The exact positions of both endoscopes were controlled radiologically. A fine scar and a channel formed around the thread. The thread facilitated use of the olive technique as well as bougienage. The olives had to be used several times, and the earlier treatment was started the sooner success was achieved (Figs. 3–6).

Since we proceeded very cautiously, treatment required several months. In both cases residual esophageal stenoses rendered subsequent bougienage necessary. We had to resect the stenoses after 10 months and 2 years respectively, and performed secondary anastomoses.

In two further cases (nos. 3 and 4) resulting moderate stenoses were treated by bougienage, initially performed at short intervals and later twice a year. All six children have developed normally without further stenosis. Figure 7 is the esophageal radiograph of case no. 4 (RD), 3½ years following elongation and operation.

Cases no. 2 and 4 later developed gastroesophageal reflux. While in the former case no symptoms were present, severe clinical signs necessitated operative treatment in the latter case, 3½ years following primary esophageal anastomosis.

Long-gap esophageal atresia is a challenge to pediatric surgeons, but prognosis is generally good. In our opinion, and according to our experience, plastic colonic replacements are no longer necessary. The death in our series was not related to the problems of long-gap atresia.



4 5 6

Fig. 4. Focusing of the pouches by means of the endoscopes

Fig. 5. Pouch-stretching and advancement of needle and thread from lower to upper pouch

Fig. 6. Thread bridges a small distance in the mediastinum

We now prefer the combination of elongation according to Howard and Myers (1965) and subsequent primary anastomosis, which has the lowest complication rate.

Endoscopic thread insertion and use of olives according to Rehbein also provided good results but caused more severe stenoses. In our opinion, this method should be reserved for longer gaps. Better results might also be achieved with this method if it were applied earlier.

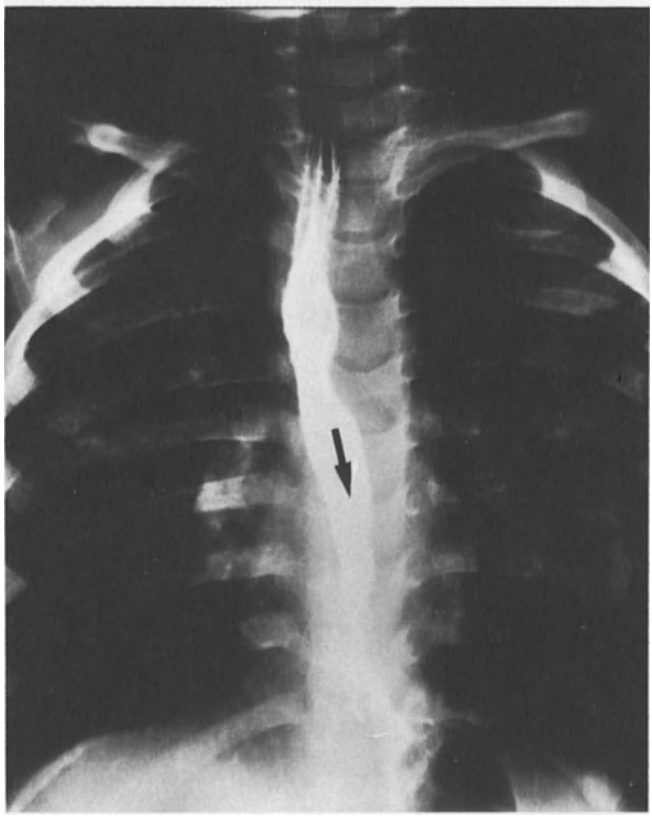


Fig. 7. Esophageal passage in patient no. 4, 3½ years after elongation and anastomosis of the esophagus

Summary

Elongation procedures for long-gap esophageal atresia are discussed. Elongation treatment according to Howard and Myers was performed in seven cases. Primary anastomosis following advance treatment could be carried out five times and esophageal elongation and anastomosis by means of the olive-and-thread method in two cases. Resulting stenoses were dilated. Endoscopic thread insertion and olive bougienage resulted in severe stenosis in two cases. This method should therefore be reserved for longer distances.

Résumé

Les techniques d'allongement de l'œsophage dans les atrésies comportant un écart important des deux extrémités, sont discutées. Dans 7 cas, on pratiqua l'allonge-

ment d'après la technique de Howard et Myers, et dans 5 cas on put faire une anastomose primitive des culs-de-sac après rapprochement; 2 fois l'élongation de l'œsophage fut pratiquée avec des olives métalliques sur un fil de Perlon, ce qui a comporté une anastomose non sanglante. Les sténoses résultantes furent dilatées. La mise en place endoscopique de fil de Perlon réalisant une anastomose non sanglante conduisit, 2 fois, à une sévère sténose, de telle sorte que cette technique doit être réservée aux cas comportant un écart considérable entre les deux culs-de-sac.

Zusammenfassung

Die Elongationsverfahren des Ösophagus bei langstreckiger Ösophagusatresie werden diskutiert. In 7 Fällen wurde die Elongationsbehandlung nach Howard u. Myers durchgeführt, 5mal konnte die primäre Anastomosierung der Blindsäcke nach entsprechender Annäherung vorgenommen werden, 2mal wurde die Ösophaguselongierung mit Metalloliven über einen Perlonfaden und unblutiger Anastomosierung vorgenommen. Resultierende Stenosen wurden bougiert. Die endoskopische Fadenlegung und Bougierung über Oliven führte 2mal zu stärkeren Stenosen und sollte daher nur für langstreckige Distanzen reserviert bleiben.

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Long-gap Esophageal Atresia: Experience with Kato's Instrumental Anastomosis, with Cervicothoracic Procedure and Primary Anastomosis, and with Retrosternal Colonic Interposition

W. Ch. Hecker[1]

The best esophagus is the esophagus which consists of tissue prepared for its function in the site assigned for it. The way to achieve this in long-gap esophageal atresia was shown by Fritz Rehbein, who developed the thread method and later the olive-thread adaptive technique. Further development led to instrumental insertion of the thread. Kato and Hollmann built a specially designed instrument for this purpose in our experimental laboratory (Kato et al. 1980). The instrument consists of a frame and two hollow bows which can be adjusted so that their tips exactly touch one another (Fig. 1). The anesthetized infant is positioned with overstretched head. Then the upper bow is led through the oral cavity to the upper pouch, the lower bow via a gastrostoma to the lower pouch (Fig. 2). When the frame is adjusted it is tested by X-ray to see whether both bows are in exactly opposite positions. Then a strong wire is brought into the lumen of the lower bow, bridging the gap and exiting with a thread attached via the lumen of the upper bow. Bougienage can now be started or, in case of a long gap, stretching dilatation using Rehbein's olive technique.

We have treated seven patients by this method, four of whom are alive and doing well. One child died of sepsis, the second of purulent mediastinitis and metastatic cerebral abscesses, and the third of a mycotic aneurysm following instrumental thread insertion. Death was caused by the method itself in the last two cases, thus demonstrating its limits. We now perform instrumental thread insertion only if both pouches are exactly adapted to each other. Thus, Kato's procedure is also suited in secondary shrinking anastomosis following primary repair if a small ureteral catheter cannot be passed over the stenosis. We have treated one child in this way.

When we recognized the limits of instrumental thread insertion for long-gap esophageal atresia, we looked for an alternative and improved the method of cervicothoracic mobilization of the esophageal pouches which we had already employed and reported on in 1972. A gastrostomy is performed immediately after establishment of the diagnosis of esophageal atresia, Vogt-type II. Two to six weeks later, when the baby is thriving or, in case of prematurity, has reached a weight of 2500 g, the definitive operation is carried out. First, the cervical esophagus is prepared via a cervical approach at the medial edge of the sternocleidomastoid muscle, whereby large vessels, the vagal nerve, and the thyroid gland are displaced to the middle. By means of a dissection swab the upper esophageal pouch

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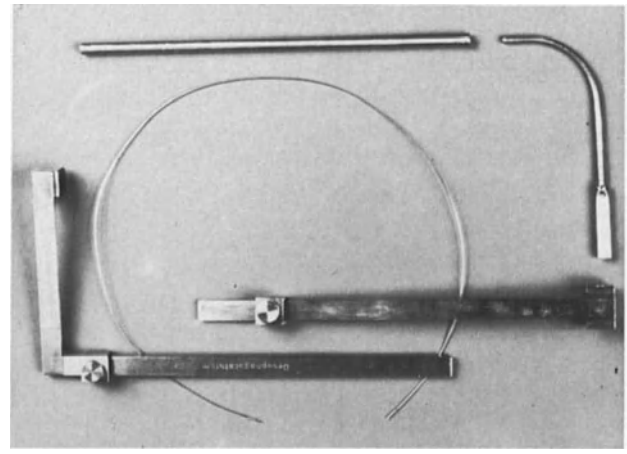

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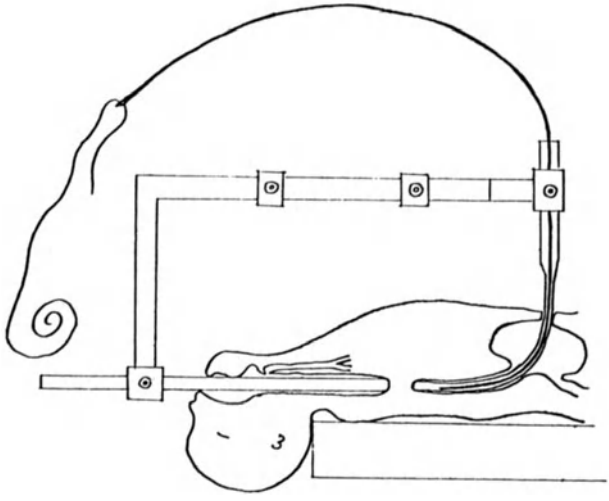


Fig. 1a, b. Kato's instrument **a** disassembled; **b** assembled and in situ

is released from its surrounding tissue and brought to the exterior. The upper pouch is then prepared up to the hypopharynx. A holding suture is then attached to the end of the upper pouch and after reposition is advanced into the mediastinum. The cervical wound is then closed, the patient is brought into a left lateral position, and a right-sided thoracotomy through the 6th intercostal space is performed. The mediastinum is exposed extrapleurally. The holding suture of the upper pouch previously advanced into the mediastinum is searched for. The upper pouch, mobilized up to the hypopharynx, can easily be drawn into the mediastinum by means of this holding suture.

Next the distal pouch is exposed; this is easily done if a Hegar dilator or a gastric tube was inserted via gastrostomy before. Another holding suture is at-

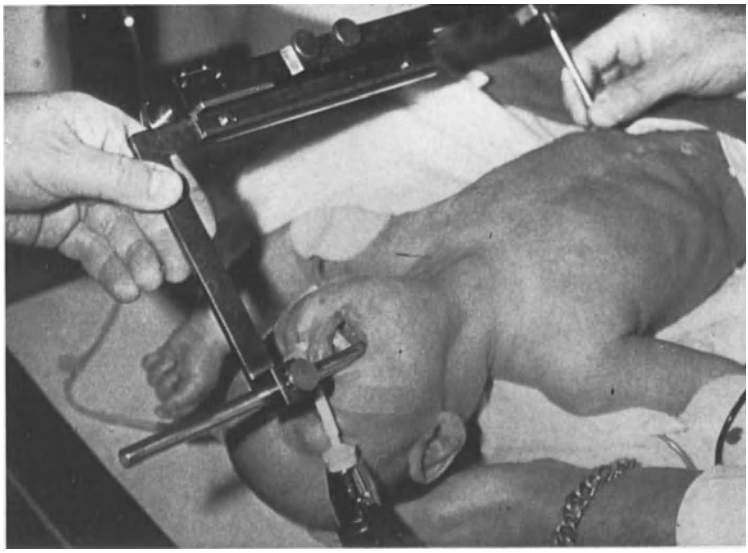


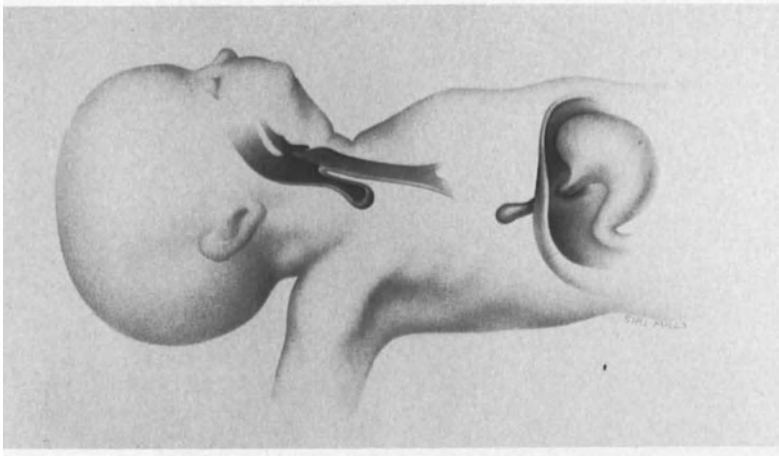
Fig. 2. Kato's instrument being used on a 4-week-old with long-gap esophageal atresia

tached to the end of the lower pouch and the pouch is prepared down to the cardia through the esophageal hiatus (Fig. 3a–e). Subsequently, both pouches are adapted to each other and primary anastomosis can be carried out if tension is low and vascularization is good. If primary anastomosis cannot be performed, the pouches remain adapted only, and anastomosis is performed 6 weeks later by means of the Kato instrument.

In another case we employed necrosing suture according to Schüllinger and Santulli (Schüllinger et al. 1982), whereby a suture through the vertices of the adapted pouches is tied tightly, thus effecting necrosis. After adaptation we inserted in addition ureteral catheter into the upper pouch and advanced its mandrin 1.5–2 cm beyond the adaptation region into the lower pouch, following air inflation of the stomach and the lower pouch. Catheter and mandrin were left in place for 4 days (Fig. 4c). On the sixth postoperative day, methylene blue, which we gave orally, appeared in the gastrostoma, thus demonstrating the creation of a tiny channel between the upper and the lower pouch. The ureteral catheter could now easily be further advanced to the stomach, taken up via the gastrostoma under radiographic control, supplied with a strong synthetic thread, and drawn back into the oral cavity. After another 10 days bougienage was started. A few months later the anastomosis was the size of a little finger, enabling the infant to feed normally (Fig. 4e).

Like Schüllinger and Santulli (Schüllinger et al. 1982), we observed cardiac insufficiency in three of our four patients treated by cervicothoracic mobilization, and retroesophageal hiatalplasty was necessary.

By means of the described method of total cervicothoracic mobilization of the esophageal pouches in long-gap atresia, it is possible in most instances to link the



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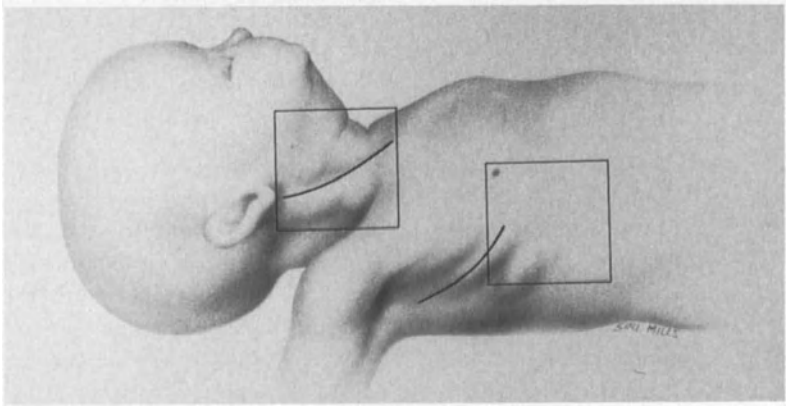

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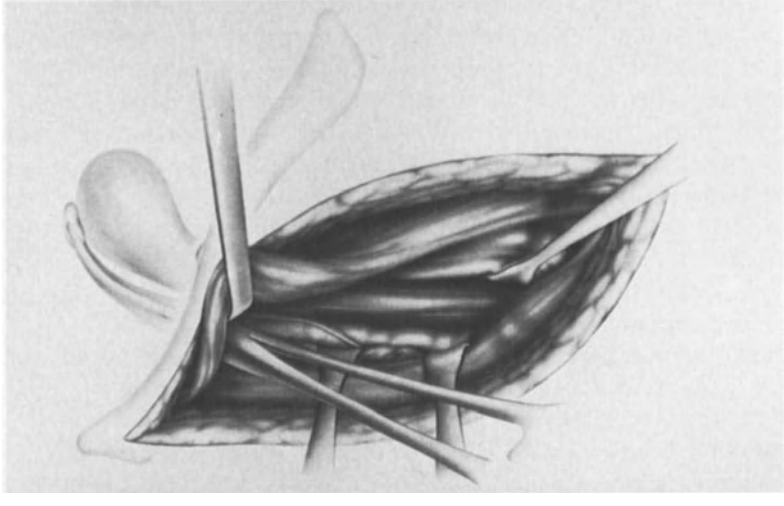

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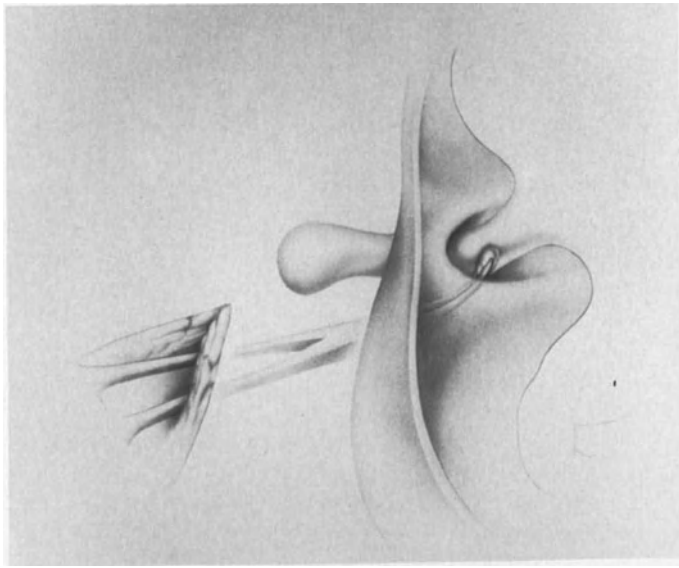

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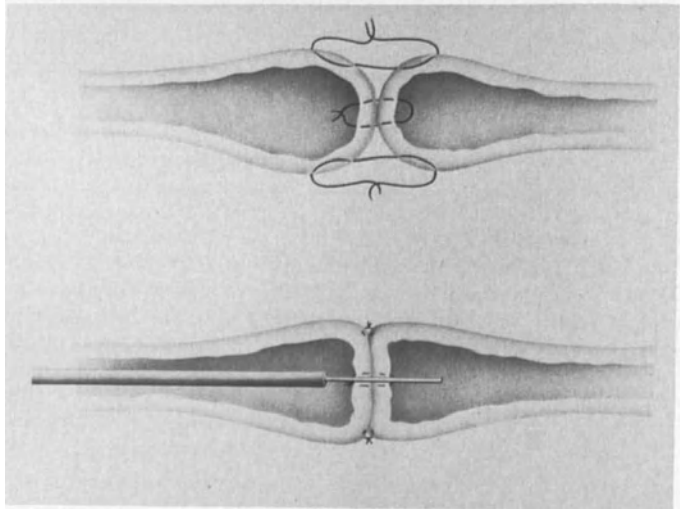

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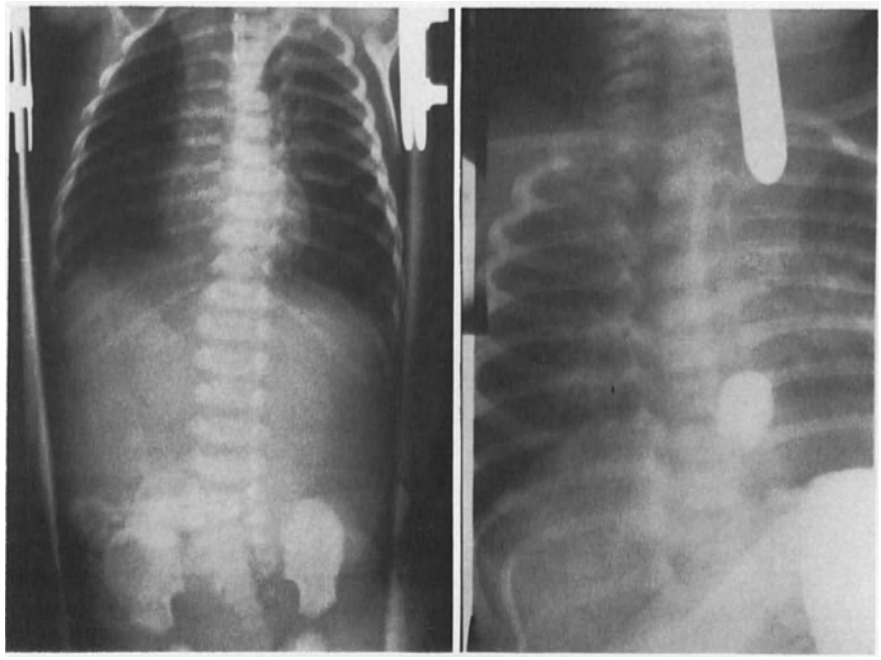
Fig. 3a–e. Our own cervicothoracic operative procedure for correction of long-gap esophageal atresia. **a** Anatomical situation of an esophageal atresia, Vogt-type II; **b** cervical incision along sternocleïdomastoid muscle, thoracic incision in the bed of the 6th rib; **c** mobilization of the upper pouch via cervical incision; **d** mobilization of the lower pouch via thoracic incision – lower pouch and cardia can be easily mobilized by means of a dissection swab; **e** *upper figure:* both pouches are adapted and are grasped by a suture through all layers of the esophageal wall, which is tied strongly so that necrosis ensues; *lower figure:* ureteral catheter with metal mandrin is passed through the upper pouch, and the metal mandrin is then advanced 1.5–2 cm into the lower pouch



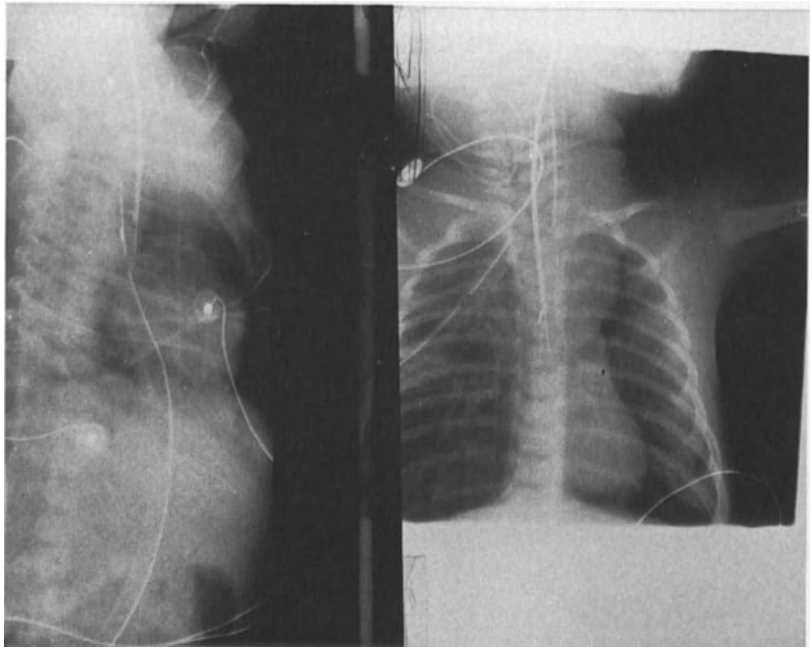
a b

Fig. 4a–f. Example of procedure presented in Fig. 3. **a** Plain abdominal X-ray – opaque gastric tube in the upper pouch filled with air; no air in the abdomen. **b** Exhibition of the lower pouch via gastrostomy at the age of 4 weeks; exhibition of the upper pouch by inserted Hegar dilator showing a distance of six intercostal spaces between upper and lower pouch. **c** X-ray immediately following operation: adaptation of both pouches, insertion of the ureteral catheter, and advancement of the metal mandrin. Adaptation area is marked by a metal clip. End of metal mandrin 2 cm distal to the ureteral catheter can be clearly seen in the lower pouch. **d** Esophageal radiogram 2 weeks following surgery; spontaneous connection of upper and lower pouch shown by contrast medium in the lower pouch. **e** Three weeks later, after first bougienage; the infant can be fed orally. Immediate and sufficient filling of the lower segment with contrast medium. **f** Eight weeks after induction of dilatation treatment; broad connection between upper and lower segment. Marked cardiac insufficiency corrected 4 weeks later. *Upper right:* good pyloric and duodenal passage

pouches either by primary anastomosis or by secondary autoanastomosis with necrosing sutures and mandrin perforation or with the Kato instrument.

If in extreme cases (Engert 1984; Maier 1985) the lower pouch is rudimentary, the procedure according to Engert may be applied: extended stretching bougienage of the upper pouch for several weeks and anastomosis, with the gastric fundus advanced into the thorax through the esophageal hiatus via the abdominal approach.

It may be helpful to perform elongation, as described by Howard and Myers and by ourselves in 1965, prior to cervicothoracic mobilization. However, a small number of patients with long-gap esophageal atresia remain, in whom the methods described cannot be employed for reasons we mention below. Esophageal



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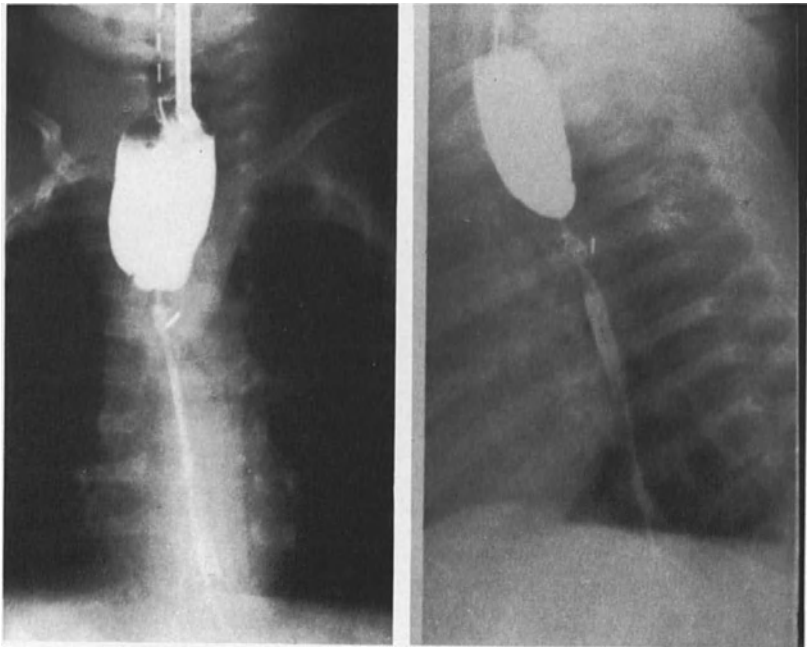

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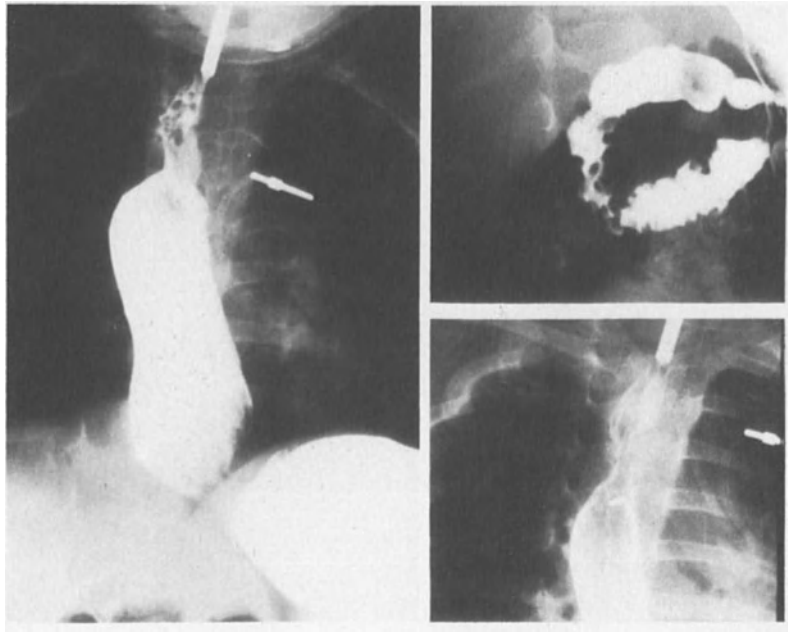

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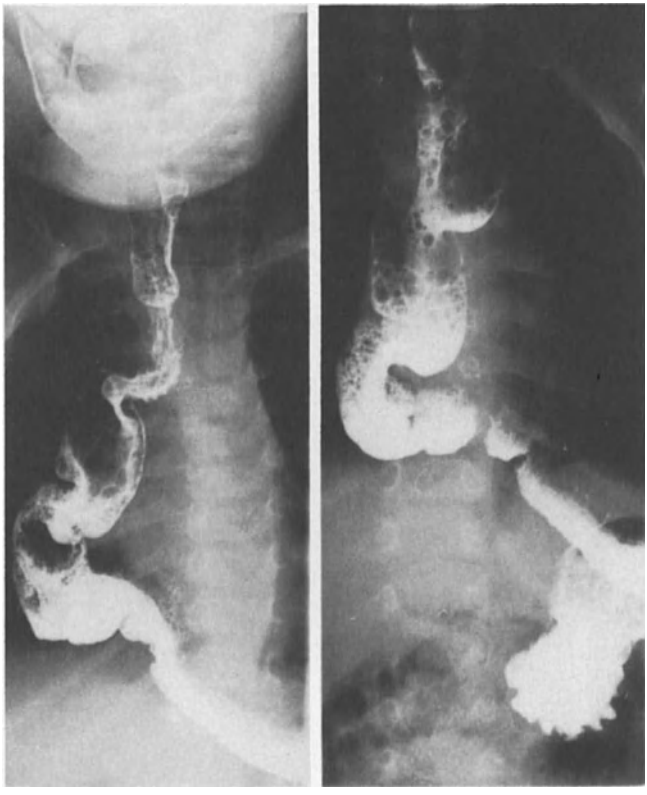

Fig. 5. Nine-month-old boy with colonic interposition between upper esophageal segment and stomach in long-gap esophageal atresia

replacement – which is the method of choice in these cases – must provide maximum security for the patient, the transplant must follow the growth of the patient, and the procedure must have minimal late complications. In our opinion, this is best achieved by colonic interposition.

Colonic interposition originated from the ideas and experimental studies of V. von Hacker, who tried an antethoracic procedure in 1894 (von Hacker and Lotheissen 1926). In 1904, Wulstein suggested intrathoracic colonic interposition (Wulstein 1910). In 1951 Camara-Lopes and Orsoni were the first to perform intrathoracic colonic interposition for esophageal replacement (Camara-Lopes 1953; Orsoni 1951). Waterston used this method from 1951 on (Waterston 1960). We (Linder and Hecker) first performed retrosternal colonic interposition in 1960 for the treatment of long-gap esophageal atresia (Linder and Hecker 1962). We now have experience with 13 patients who underwent colonic interposition. There was no surgical mortality; one child died at home of pneumonia 1 year following surgery (Fig. 5).

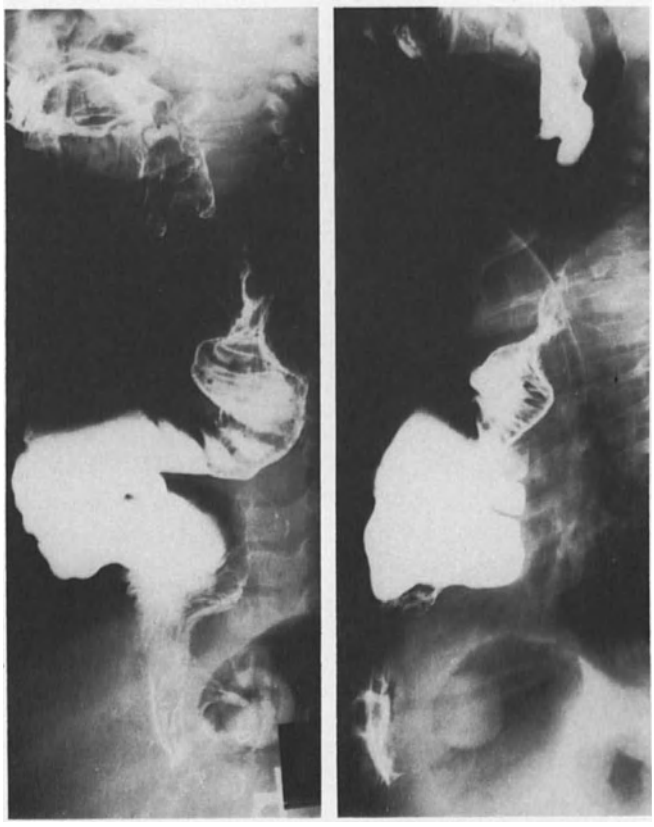

Fig. 6. Child with retrosternal colonic interposition in long-gap esophageal atresia. Due to mediastinal hernia interposed colon is displaced into the right thoracic cavity; free passage

Among the organs under discussion (stomach, small bowel) the colon is in our opinion best suited for esophageal replacement, since it can be securely mobilized due to its marginal vessel arcades. The retrosternal procedure is the safest, since only one large body cavity must be opened; gastrocolonic anastomosis is more reliable than intrathoracic anastomosis between the colon and the serosaless lower esophageal pouch. Moreover, the supplying blood vessel is less endangered than it is in the left-sided intrathoracic procedure according to Waterston (1960), in which the blood vessels are positioned within diaphragmatic incisions.

We observed the following late complications in our 13 patients from 1960 to 1983: in three patients no pyloroplasty had been performed, as we do routinely today, and we had to make up for this several years later. One patient had to undergo relaparotomy for stenosis following pyloroplasty.

Reflux colitis occurred in a patient in whom the colon had to be interposed anisoperistaltically for technical reasons, and where no primary pyloroplasty had been performed. Initially, the stenosis could be treated by bougienage; 15 years

later, however, stenosis became so severe that a longitudinal incision and transverse suture combined with a pyloroplasty were necessary. This was 10 years ago, and the patient has been free of complaints since then.

In another patient a stenosis of the colon at the level of the diaphragm required dilatation 3 years following colonic interposition. Another child developed a mediastinal hernia of the interposed colon into the right pleural cavity. About one half of the colon is not in the retrosternal position but runs loopshaped, intrapleurally. The passage, however, is free 14 years after surgery (Fig. 6).

Dilatation according to Heinike and von Mikulicz for stenosis of the cervical esophagocolonic anastomosis had to be carried out in another instance, 2 years following surgery. Two children developed pigeon breast due to an overgrowth of the left costochondral junctions above the interposed colon. Surgical correction was not necessary, however.

A child who had undergone colonic interposition in another hospital according to Waterston's technique (Waterston 1960) had a caudal loop of the lower esophagus causing kinking and stenosis at the lower esophagocolonic junction. This stenosis was operated on by means of a longitudinal incision and transverse suturing and the caudal elongation of the interposed colon was done with purse-string sutures. The complications described here could be mastered and disappeared following surgical correction.

Recently, colonic interposition was indicated in three children with long-gap esophageal atresia where elongation treatment (which is usually the method of choice in these cases) could not be employed. In two patients purulent mediastinitis following recurrence of esophagotracheal fistulas led to necrosis of the primary esophageal anastomosis. After necrosectomy the distal esophageal segment was closed blindly and the proximal segment was led to the exterior via a cervical incision. The lengths of the proximal and distal segments did not allow for another direct anastomosis. Almost complete shrinking of the distal esophageal segment was disclosed radiologically in both cases.

The third baby had, besides long-gap esophageal atresia, aplasia of the right lung and a malformation of the large vessels. The upper pouch ended 1 cm above the jugulum. After dissection of the fistula between the tracheal bifurcation and the lower esophageal segment and fistulation of the upper, extremely short pouch, the baby developed tracheal stenosis caused by the vascular malformation, which required aortotruncopexy to the sternum. Another left-sided thoracotomy seemed to be useless in this case and colonic interposition was performed.

Summary

Twenty-four patients with long-gap esophageal atresia have been treated since 1960. Instrumental anastomosis according to Kato was applied in seven cases, cervicothoracic procedure with primary anastomosis followed extended mobilization of the upper and lower pouches in four cases, and retrosternal colonic interposition was performed in 13. Currently, treatment of long-gap esophageal

atresia, Vogt-type II, should include first a gastrostomy and, 4–6 weeks later, if the baby is thriving, extended cervicothoracic mobilization of the upper pouch up to the larynx and of the lower pouch down to the cardia, followed by primary anastomosis. If primary anastomosis cannot be performed, bouth pouches should be adapted, by necrosing sutures according to Santulli, and perforated with a thin mandrin. Thus, esophageal replacement is necessary only in exceptional cases, for which retrosternal colonic interposition is recommended.

Résumé

Vingt-quatre patients avec atrésies comportant une longue distance entre les culs-de-sac furent traités depuis 1960. Les anastomoses instrumentales d'après la technique de Kato furent réalisées dans 7 cas, une technique cervico-thoracique avec anastomose primaire consécutive à une mobilisation extensive de l'extrémité supérieure et de l'extrémité inférieure furent pratiquées dans 4 cas, et une interposition rétrosternale du côlon dans 13 cas.

En conséquence, le traitement des atrésies œsophagiennes "long-gap", type Vogt II, doit comporter tout d'abord une gastrostomie et ensuite, 4 à 6 semaines plus tard, lorsque l'enfant s'est bien comporté, une mobilisation cervico-thoracique extensive du cul-de-sac supérieur jusqu'au larynx et du cul-de-sac inférieur jusqu'au cardia, suivie d'une anastomose primitive. Si cette anastomose ne peut pas être pratiquée, les deux culs-de-sac doivent être rapprochés avec des sutures nécrosantes selon Santulli et perforés ultérieurement avec un mandrin étroit. De cette sorte, le remplacement de l'œsophage est exceptionnellement nécessaire. Dans ce cas, une interposition colique rétrosternale est recommandée.

Zusammenfassung

Es liegen Erfahrungen von 24 Kindern mit langstreckiger Ösophagusatresie im eigenen Krankengut seit 1960 vor. Bei 7 Patienten wurde instrumentell mit dem Kato-Gerät eine Anastomose erreicht, in 4 Fällen wurde nach vollständiger Mobilisierung des oberen und unteren Blindsacks durch zervikothorakales Vorgehen die Speiseröhrenkontinuität hergestellt, und bei 13 Patienten wurde ein Ösophagusersatz durch Kolon, welches retrosternal nach kranial geführt wurde, vorgenommen.

Die derzeitige Vorstellung ist, bei einer langstreckigen Ösophagusatresie vom Typ Vogt II zunächst eine Gastrostomie anzulegen und 4–6 Wochen später nach einwandfreiem Gedeihen des Kindes durch ausgedehnte zervikothorakale Mobilisierung des oberen Segments bis zum Larynx und des unteren Segments bis zur Kardia eine primäre Anastomose zu erreichen. Wenn dies nicht gelingt, werden die beiden Segmente adaptiert, mit der sog. durchschneidenden Naht nach Santulli versehen und mit einem Ureterenmandrin perforiert. Nur in Ausnahmefällen ist heute noch ein Ösophagusersatz notwendig. Wenn man sich dazu entschließt,

wird hier das Kolon empfohlen und der retrosternale Weg als der sicherste angesehen.

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Esophagus Replacement by Free, Autologous Jejunal Mucosa Transplantation in Long-gap Esophageal Atresia

H. Halsband[1]

Various elongation methods (see Table 1) have developed since 1965 according to N. A. T. Myers' motto: "The best esophagus is the patient's own esophagus"; they have effected a decisive change in treatment, rendering replacement methods which are burdened by a high complication rate avoidable.

However, these elongation methods themselves cannot be applied without problems. The preoperative elongation methods require repeated stretching bougienage (Howard and Myers 1965; Hays et al. 1966; Hendren and Hale 1975) or dynamic dilatation (Hofmann 1975) for several weeks. During this time there

Table 1. Methods for bridging long-gap esophageal atresias

A Elongation of the segments (with subsequent end-to-end anastomosis)
- I Stretching bougienage
  - 1 Temporary stretching of the upper pouch (Howard and Myers 1965)
  - 2 Temporary stretching of both pouches (Hays et al. 1966)
  - 3 Intermittent stretching of both pouches by application of an electromagnetic field (Hendren and Hale 1975)
- II Operative procedure
  - 1 Transverse dissection and longitudinal closure of the anterior wall of the proximal pouch (Okmian and Livaditis 1969)
  - 2 Circular myotomy of the proximal pouch (Livaditis et al. 1972)
  - 3 Cervical mobilization of the proximal pouch (Daum et al. 1970)
- III Transmediastinal thread technique, with or without olive, and postoperative "autoanastomosis" (Rehbein and Schweder 1971)
  - 1 Following endoscopical thread insertion (Okmian et al. 1975)
  - 2 Following thread insertion with the Kato instrument (Kato et al. 1980)
- IV Dynamic dilatation of the distal pouch by balloon catheter and silastic tie in the cardiac region (Hofmann 1975)

B Replacement techniques (with or without prior elongation)
- I Pedunculated colon transposition (Sandblom 1948)
- II "Gatric-tube" technique (Burrington and Stephens 1968)
- III Pedunculated small-bowel transposition (Longmire 1951)
- IV Free jejunal mucosa/submucosa interposition (Halsband 1977)

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is always danger from aspiration due to the upper pouch and from perforation during bougienage. The intraoperative methods of circular myotomy (Livaditis et al. 1972) or cervical mobilization (Daum et al. 1970) often do not provide enough length. The transmediastinal olive-and-thread method (Rehbein and Schweder 1971) may lead to mediastinitis or sometimes to a brachyesophagus with gastroesophageal reflux.

Our Method

We started from the reflection that bridging of the long gap consists primarily of only one layer of tissue both in Livaditis' myotomy (Livaditis et al. 1970) and in Rehbein's (Rehbein and Schweder 1971) olive-and-thread technique. Livaditis' myotomy uses primarily a mucosa/submucosa tube with secondary coating by fibrous tissue, Rehbein's olive-and-thread method primarily a channel of mediastinal granulation tissue with secondary esophageal mucosa lining. We therefore tested, in animal experiments (1977), whether long-gap esophageal atresias could be bridged by free-transplanted, autologous mucosa/submucosa tubes from the jejunum.

We chose the jejunum based on the investigations of Wilfingseder and coworkers (1971), since the jejunal epithelium is one of the most vital body tissues, able to regenerate entirely within 1 week. Wilfingseder et al. (1971, 1972) applied autologous, deserosized jejunal segments as free transplants for vaginal replacement in five women.

Since Wilfingseder's histological findings revealed an extensive loss of the muscularis layer, we decided to also remove the muscularis, which is very susceptible to ischemia, and to transplant merely a mucosa/submucosa tube providing for secure healing. Trophic supply, initially occurring exclusively by diffusion, is the more critical the thicker the free transplant is.

Preparing the Transplant

The separation of serosa and muscularis layers from submucosa and mucosa (Fig. 1) ensues in the outer smooth, soft, and well-vascularized submucosa, the so-called shifting layer of the bowel, which is relatively rich in elastic fibers. Furthermore, muscularis mucosae bundles penetrate into the submucosa (Bargmann 1967), whereby the union of submucosa and mucosa is maintained during preparation.

Figure 2 shows cross-cuts through the small bowel of a newborn baby and through the prepared mucosa/submucosa tube. After removal of serosa and muscularis propria, the mucosa unfolds, providing a flat and regular contact between well-vascularized submucosa and surrounding tissue. This could not be achieved by transplantation of a complete or only deserosized bowel segment (Wilfingseder et al. 1972; Oesch et al. 1980).

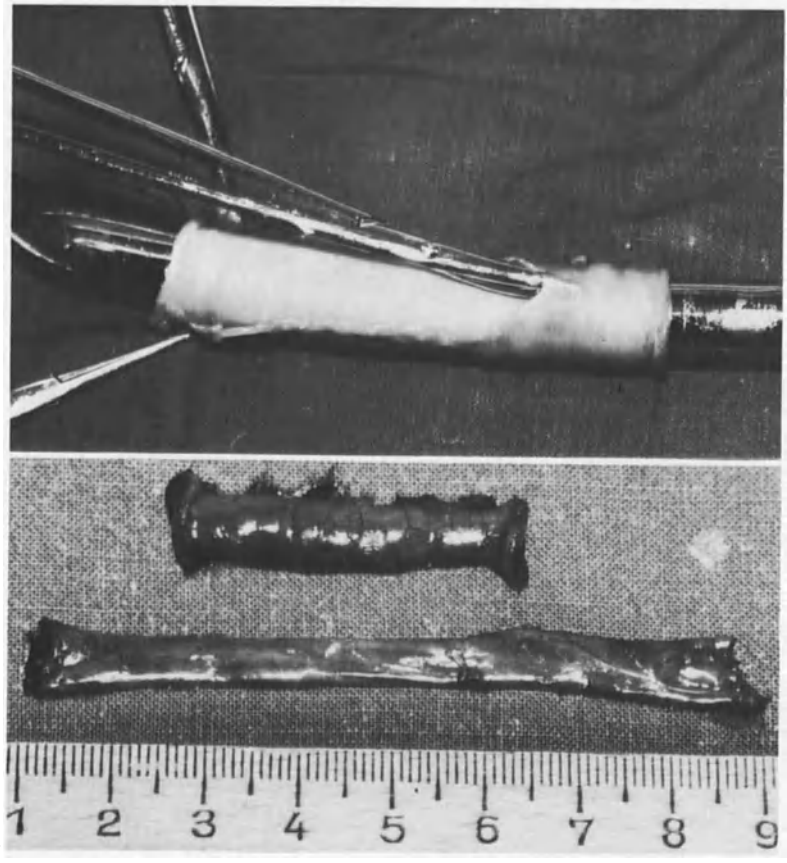


Fig. 1a, b. Preparation of the transplant. **a** Small-bowel segment drawn over a Hegar dilator; careful separation of the layers within the submucosa, dissection of serosa and muscularis. **b** Jejunal segment and prepared mucosa/submucosa tube of a newborn

Animal Experiments

Experimentally, we replaced a part of the thoracic esophagus (8 cm on average) by free-transplanted mucosa/submucosa tubes of corresponding length in 32 young beagles. The free-transplanted jejunal tubes prevented transmural contamination of the mediastinum around the esophageal defect.

Esophageal splinting for 6 weeks – beyond the 2–3 week period of the most intense wound contraction – and bougienage immediately following surgery confined circular transplant shrinking and avoided stenosis as well as longitudinal shrinking. The transplants in the dogs lost their shrinking tendency at 3–4 months postoperatively. Figure 3 shows esophageal radiograms of a dog, 6 weeks and 2 years following 8-cm esophageal replacement.

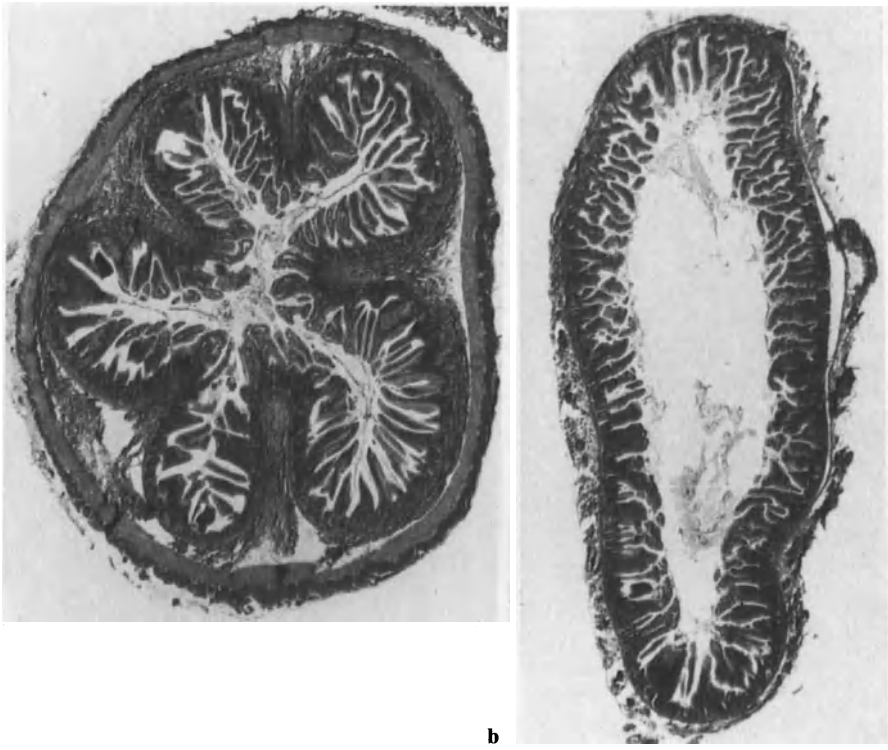


Fig. 2. a Cross-section through the jejunum of a newborn, × 13. **b** Cross-section after removal of serosa and muscularis propria; the mucosa tube has unfolded, × 13

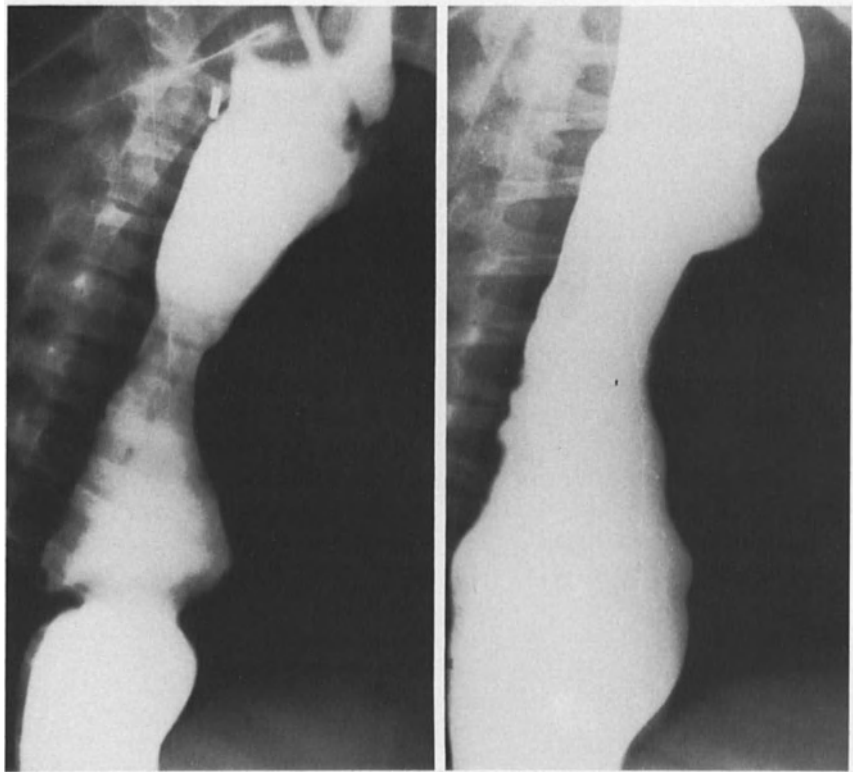
Histological evaluation revealed healing of the free-interposed autologous jejunal mucosa transplants by formation of granulation tissue tubes together with the mediastinal wound bed, lined partly by persistent jejunal epithelium and partly by ingrowing esophageal epithelium. The dogs were followed up for 6 years. There was no substantial dysphagia in the long-term surviving animals.

Clinical Application

From 1977 to 1982 free jejunal mucosa interposition was applied in two of five cases of long-gap esophageal atresia.

Operative Procedure

In suspected long-gap esophageal atresia, the newborn is brought into a position which allows the approach to both large body cavities. First, a thoracotomy is performed, clarifying the anatomical situation and confirming the long-gap. Then the



a b

Fig. 3a, b. Esophagograms of a dog following 8-cm esophageal replacement by jejunal transplantation. **a** Six weeks postoperatively, anastomotic region marked by cording rings and clips; **b** 2 years postoperatively

thoracic retractor is removed and the thoracic wound covered with a wet swab. This is followed by laparotomy, resection of an upper jejunal segment of 10 cm in length, end-to-end anastomosis, and preparation of the graft.

For graft preparation, the jejunal segment is drawn over a Hegar dilator (Fig. 1a). Serosa and muscularis propria are cut through to the submucosa at one border, and then the layers are separated within the submucosa. Thereafter, serosa and muscularis are longitudinally dissected and removed in toto. The prepared mucosal tube has nearly double the length of the original jejunal segment (Fig. 1b). The laparotomy wound is also covered with a wet swab during preparation and the thoracic operation is then continued.

Interposition of the transplant is performed under slight tension by means of two anastomoses. We recommend fixation of both esophageal pouches to the surrounding mediastinum in order to reduce tension to the anastomoses and to avoid healing and diffusion disturbances of the graft by esophageal peristalsis.

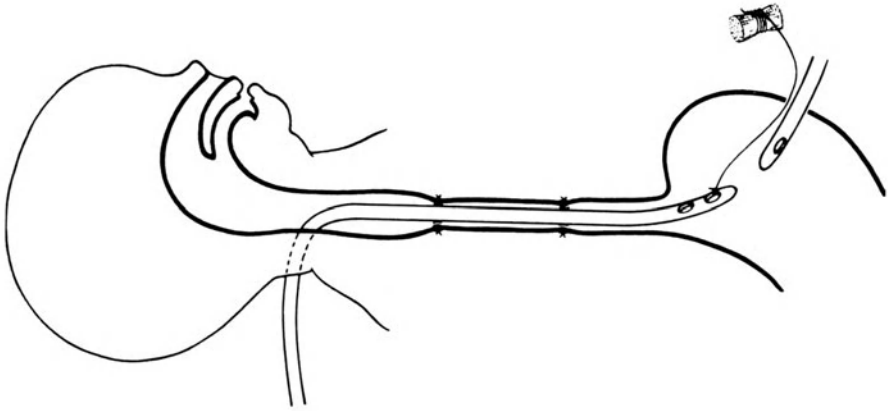


Fig. 4. Splinting of the transplant by a thick gastric tube running to the exterior through a cervical esophagostoma. Distal end is connected to a thread running through gastrostoma additionally created

We insert a 12-Charrière gastric tube for splinting and feeding for 4–6 weeks, i.e., beyond the period of the most intense wound contraction (Fig. 4).

Proximally, the splinting tube runs to the exterior through a cervical esophagostoma or a pharyngostoma according to the "tube-pharyngostomy" technique suggested by Talbert and Haller (1965), thus avoiding decubital necrosis of the larynx. The distal border of the splinting tube is connected to a synthetic thread running to the exterior through a gastrostoma which is also created.

Case 1

Newborn baby, birth weight 2900 g, long-gap esophageal atresia type IIIb according to Vogt, with hypoplastic lower pouch (Fig. 5a). After extended mobilization of both pouches, a free jejunal mucosa/submucosa transplant of 5 cm in length was interposed. Splinting for 4 weeks followed in the above-described manner. Esophageal radiograms at 4 weeks (Fig. 5b) and 10 weeks (Fig. 5c) following operation showed free passage, with no evidence of fistula.

The child also had severe shunting due to congenital heart disease, which was, however, diagnosed only postoperatively. This malformation required pulmonary banding performed at the *Medizinische Hochschule Hannover*. Postoperative suction treatment caused recurrence of the esophagotracheal fistula, necessitating rethoracotomy and closure of the fistula. The child died 3 months later from the effects of congenital heart disease and pneumonia.

Postmortem examination (Fig. 6) showed an intact and unshortened transplant, regular anastomoses, no shrinking, and no fistula. There was a strong cicatricial coating of the transplant and complete healing of the jejunal mucosa tube. Histological examination[2] (Fig. 7) showed not only persistence of the entire

[2] Histological findings courtesy of Dr. Völpel, then at the Institute of Pathology, *Medizinische Hochschule Lübeck*

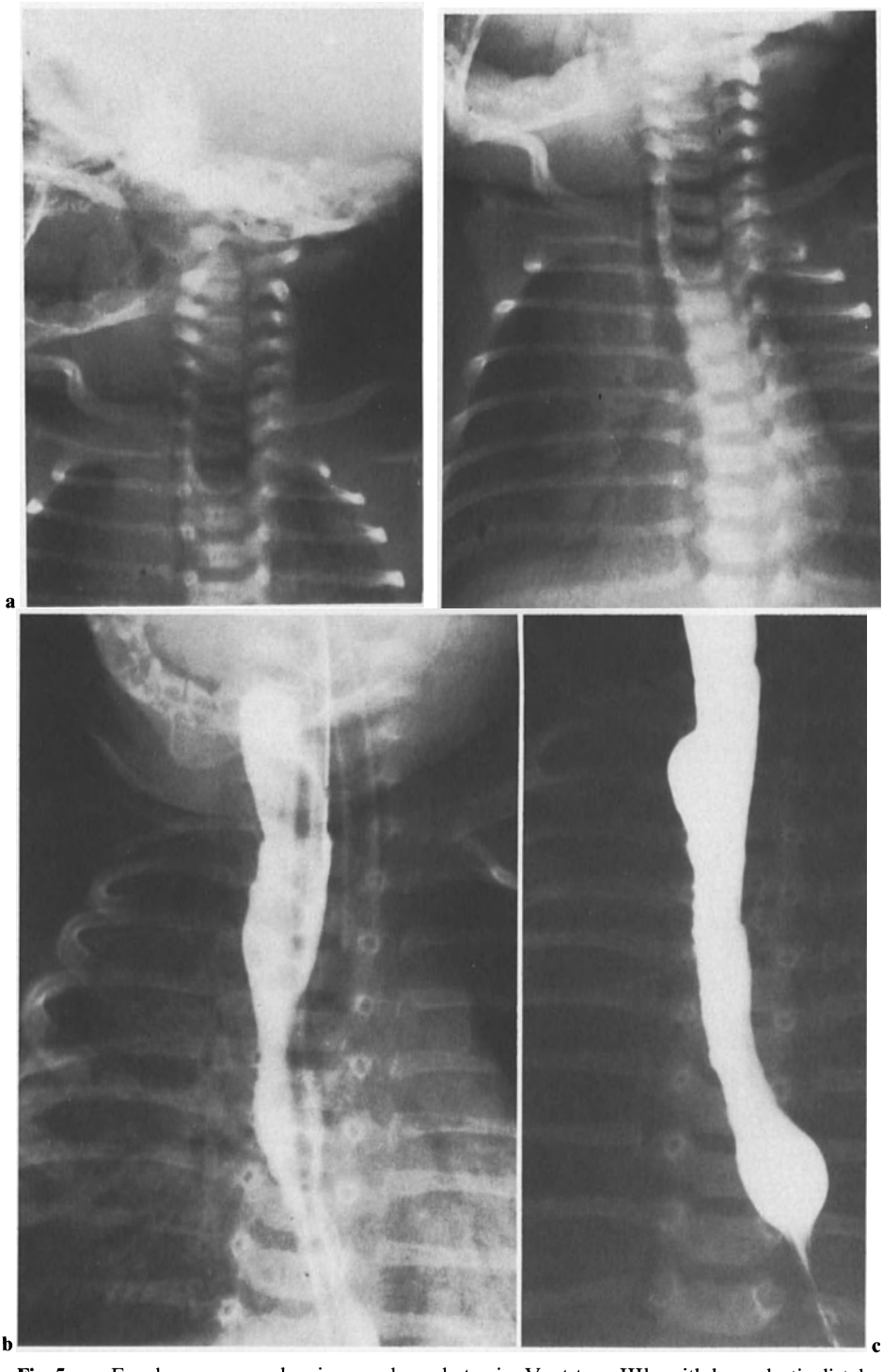


Fig. 5a–c. Esophagograms **a** showing esophageal atresia, Vogt-type IIIb, with hypoplastic distal pouch and long gap; **b** 4 weeks following interposition of a 5-cm jejunal mucosa transplant; **c** 10 weeks postoperatively

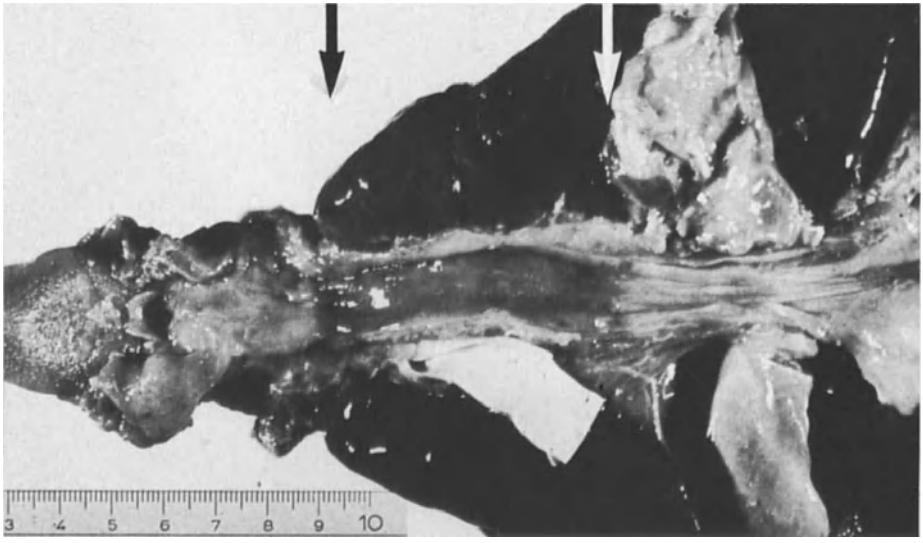
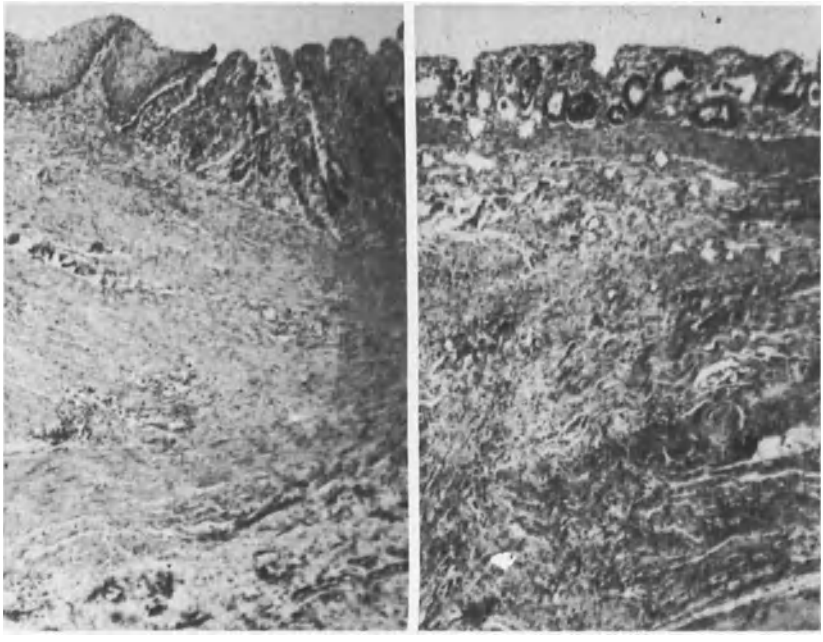
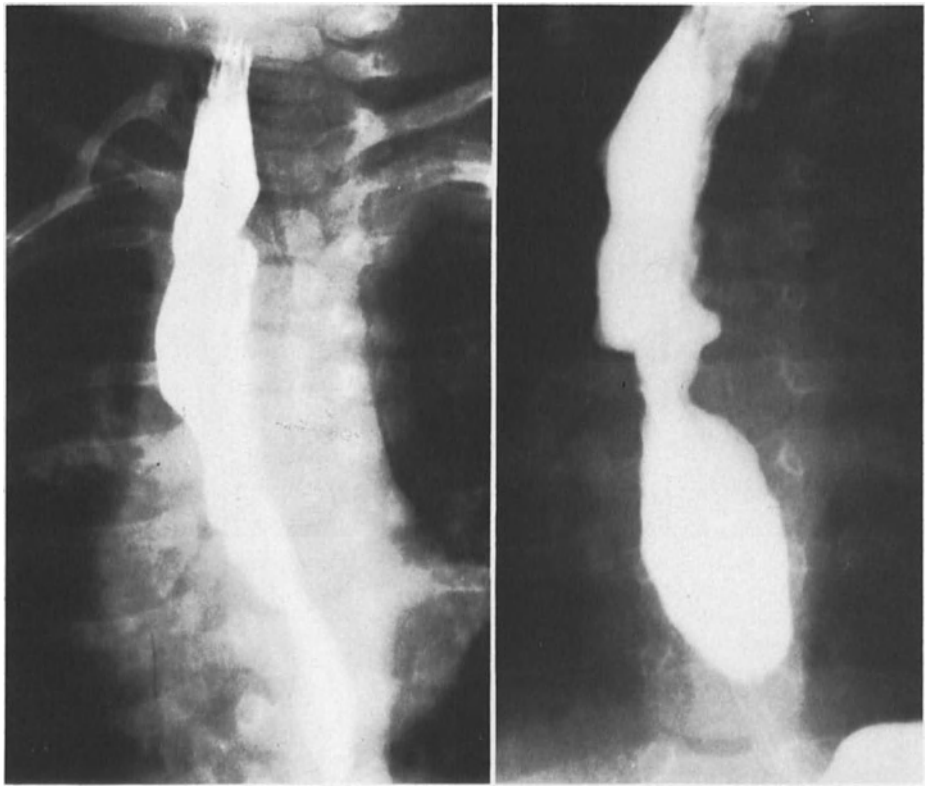


Fig. 6. Autopsy preparation 3 months postoperatively: 5-cm transplant between the esophageal segments (upper segment, 3 cm; lower segment, 4 cm) is unshortened; regular anastomoses, strong cicatricial coating of the completely healed transplant. *Arrows* point at the upper and lower anastomosis



a **b**

Fig. 7a, b. Histological examination showing **a** proximal esophagus and transition into transplant with persistent jejunal mucosa; dense, fibrotic scar, poor in cells, *below;* **b** mid region of transplant. HE; × ca. 35



a b

Fig. 8a, b. Esophagograms made **a** 3 months following interposition of a 5-cm jejunal mucosa transplant, and **b** 7 months postoperatively

mucosa but also persistence of many muscularis mucosae fibers. The surrounding cicatricial tissue could be clearly seen.

Case 2

Newborn baby, birth weight 2750 g, suspected Holt-Oram syndrome with multiple malformations: scoliosis of the thoracic column, rib dysplasias, dextrocardia, hypoplasia of the left radius and aplasia of the right radius, severe hand malformation, hypoplasia of both thumbs, and flexion contraction of the fingers. The anatomical situs of the esophagus was the same as in case 1.

We interposed a 5-cm long free jejunal transplant in the same way; cervical esophagostomy and gastrostomy were also performed. The postoperative course was at first uneventful. Two weeks later, the infant developed bilateral pneumonia with protracted healing, followed by recurrent atelectasis and bronchopneumonic infiltration. In the esophagograms obtained 3 months (Fig. 8a) and 7 months (Fig. 8b)

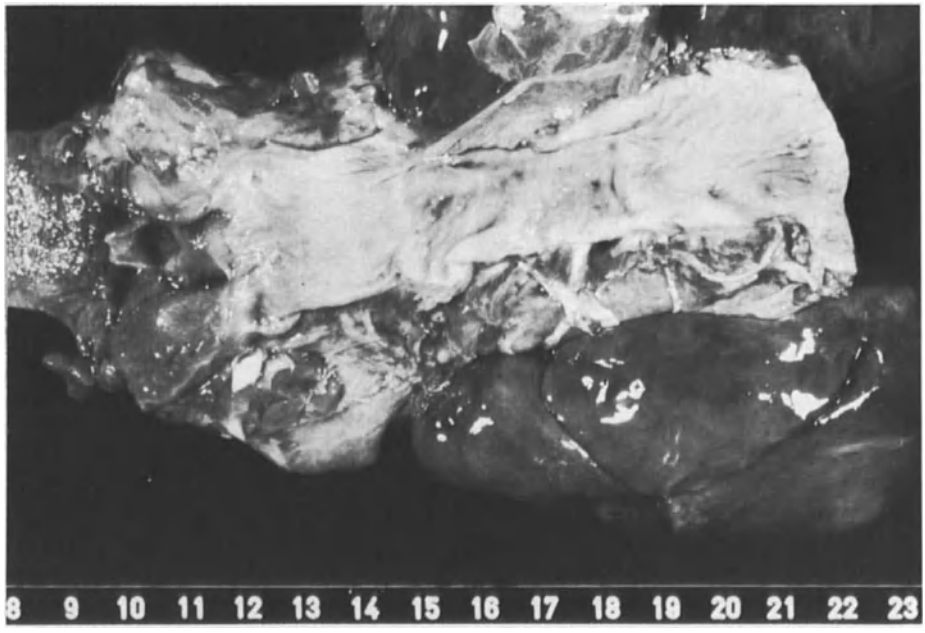


Fig. 9. Autopsy preparation 1 year postoperatively, showing proximal esophageal segment (3.5 cm), transplant (5 cm), and distal segment (4.5 cm); almost entirely preserved jejunal mucosa

postoperatively free passage without leakage could be demonstrated, associated, however, with gastroesophageal reflux.

At home the child again developed pneumonia, and he died 1 year following the operation. Postmortem examination revealed marked distention of both cardiac ventricles with no signs of cardiac malformation, as well as bilateral suppurative pneumonia. The autopsy preparation (Fig. 9) shows the esophagus with its proximal segment (3.5 cm), transplant (5 cm), and distal segment (4.5 cm), no fistula, and almost entirely preserved jejunal mucosa. Histologically, a transition zone from esophageal squamous epithelium to jejunal mucosa can be seen (Fig. 10a) within the anastomotic region. The muscularis mucosae layer is hyperplastic. There is persistent villose jejunal mucosa in the mid region of the transplant (Fig. 10b) and slight fibrosis of the submucosa.

Discussion

Although we had the opportunity only twice to apply free jejunal mucosa interposition in newborns with long-gap esophageal atresia, and cannot report on long-term results owing to the death of the patients from severe concomitant malformations, our experimental and clinical experience seems to justify the recommen-

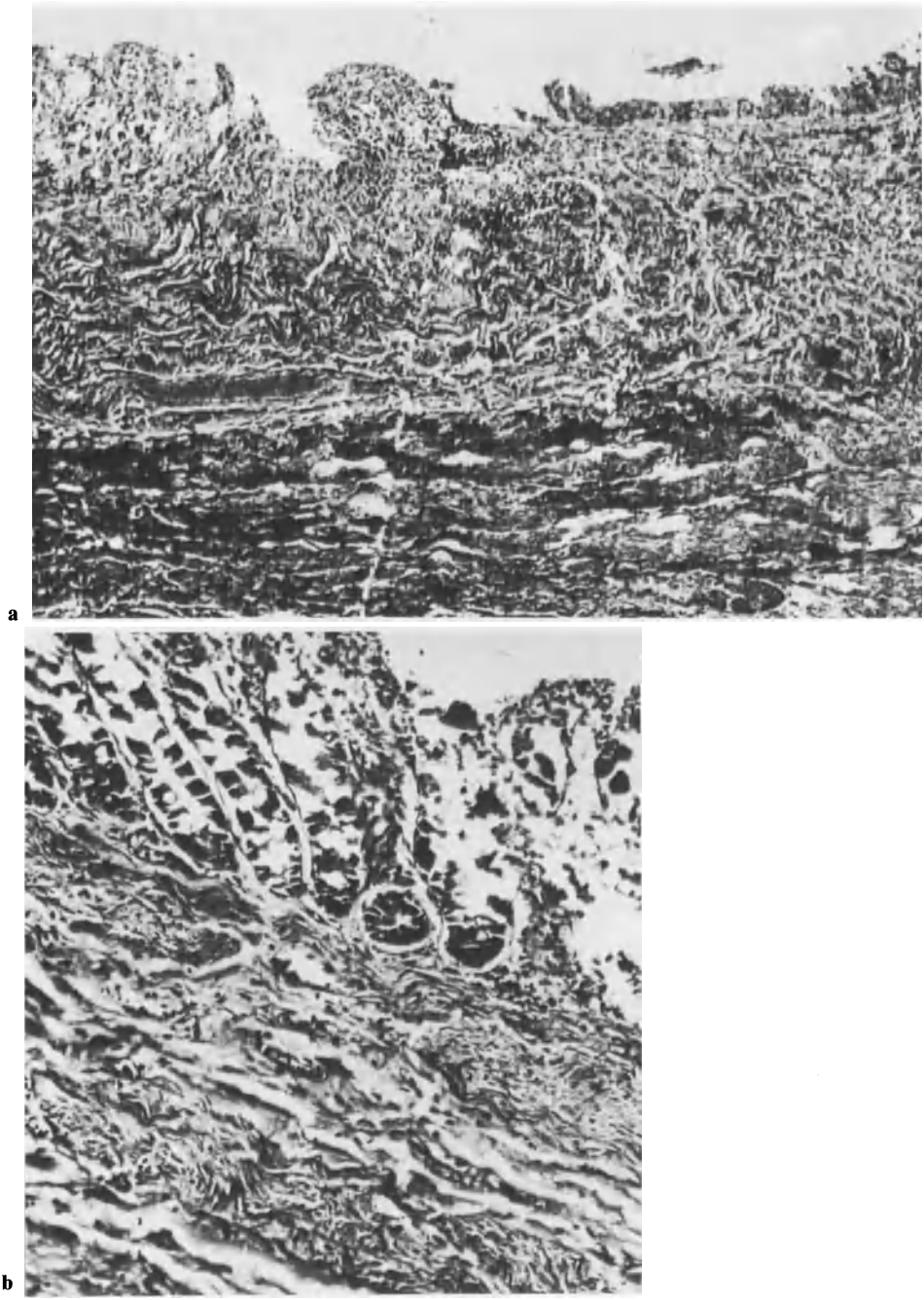


Fig. 10a, b. Histological findings in case 2, 1 year postoperatively. HE; × ca. 140. **a** Proximal anastomotic region: transition zone from esophageal squamous epithelium to small-bowel mucosa *(left side)*. **b** Mid region of transplant showing villose structure of the jejunal mucosa, slight fibrosis in the submucosa

dation of this procedure as an alternative to the techniques commonly employed in the treatment of long-gap esophageal atresia.

The death of our two children, however, gave us the opportunity to evaluate the operative results thoroughly and to confirm the results we had achieved in preceding animal experiments.

Generally, total correction in one session seems to be desirable and practicable in mature newborns without severe associated malformations. In our two patients, laparotomy and small-bowel resection extended the time of operation by approximately 45 min. The advantages of free jejunal mucosa/submucosa interposition can be summarized as follows:

- Primary, definitive bridging of the long-gap is possible.
- Compared with elongation techniques
  a) risks from the upper pouch can be avoided,
  b) longer gaps can be bridged,
  c) there is less risk with regard to mediastinitis and brachyesophagus.
- Compared with replacement techniques using pedunculated gastric or small-bowel segments and requiring repeated sessions there is less risk.
- Time of hospitalization can be shortened considerably.

Summary

There is apparently no ideal operative technique in the treatment of long-gap esophageal atresia, as is shown by the plurality of operative procedures described in the literature. Our own technique is presented, based on vaginal replacement by free, deserosized jejunal segments according to Wilfingseder. In addition, the muscularis propria layer was removed to improve trophic supply of the transplant which occurs initially only by diffusion. The free transplanted jejunal mucosa segments prevented transmural contamination of the mediastinum within the esophageal defect. Former animal experiments on beagles, as well as clinical experience with two newborn babies with long-gap esophageal atresia, showed that splinting for 4–6 weeks – beyond the time of the most intense wound contraction – and subsequent dilatation treatment could prevent circular and longitudinal shrinking of the transplants, thus avoiding stenosis.

Histological findings revealed that on the average free-transplanted jejunal mucosa/submucosa tubes 8 cm long in dogs and 5 cm long in babies formed tubes of granulation tissue with the surrounding mediastinum, lined by persistent jejunal epithelium and partly by ingrowing esophageal epithelium. The two babies died ¼ and 1 year following the operation, from their severe associated malformations. This, however, enabled us to do a thorough morphological examination of the interposed transplants and to document the complete healing. The advantages of the method described are (a) primarily definitive bridging of the defect, (b) less problems and risks as compared with elongation methods, and (c) possibly shorter hospitalization.

Résumé

Il n'y a, apparemment, aucune solution thérapeutique idéale dans le traitement des atrésies œsophagiennes "long-gap", quand on constate le grand nombre de corrections chirurgicales décrites dans la littérature.

Une technique personnelle est présentée, basée sur le remplacement vaginal par un segment jéjunal libre dépouillé de sa séreuse, selon Wilfingseder. En complément, la couche musculaire propre a été enlevée pour améliorer la trophicité du transplant qui est nourri seulement au début par diffusion. Le transplant jéjunal libre de muqueuse évite des contaminations transmurales du médiastin que l'on constate dans les défects œsophagiens.

Les expériences sur le chien, de même que l'expérimentation clinique sur deux nouveau-nés avec atrésie œsophagienne "long-gap", ont montré que le passage d'un cathéter pendant 4 à 6 semaines, c'est-à-dire la période de rétraction la plus intense et des dilatations successives, pouvait prévenir la rétraction circulaire et longitudinale du transplant, évitant ainsi les sténoses.

Les constatations histologiques ont montré qu'un transplant de muqueuses et sous-muqueuses jéjunales libres de 8 cm de longueur chez le chien et de 5 cm chez l'enfant, occasionne la formation d'un tube de granulations au contact du médiastin environnant, bordé par un épithélium jéjunal persistant et aussi partiellement par un épithélium œsophagien invasif. Les deux enfants sont décédés 3 et 12 mois après l'opération de malformations associées sévères. Cependant, l'étude du transplant interposé a montré une cicatrisation complète.

Les avantages de la méthode sont:

1) pontage primaire définitif du défect,
2) problèmes et risques mineurs comparativement aux méthodes d'élongation,
3) possibilité de raccourcir le temps d'hospitalisation.

Zusammenfassung

Die Vielzahl der angegebenen Operationsverfahren zur Behandlung langstreckiger Ösophagusatresien beweist, daß kein Verfahren ideal ist. Ein eigenes Vorgehen wird vorgestellt, das seinen Ursprung hat in den von Wilfingseder vorgeschlagenen Vaginalersatzplastiken mit freien deserosierten Jejunumsegmenten. Um die anfänglich nur durch Diffusion erfolgende Transplantaternährung günstiger zu gestalten, wurde auch die Muscularis propria entfernt. Die frei transplantierten Jejunumschleimhautinterponate verhinderten eine transmurale Keimkontamination des Mediastinums im Ösophagusdefektbereich. Frühere tierexperimentelle Studien an jungen Beaglehunden sowie die klinischen Erfahrungen mit 2 Neugeborenen mit langstreckigen Atresien zeigten, daß durch 4- bis 6wöchige Schienung – über die Phase der intensivsten Wundkontraktion hinaus – sowie durch anschließende Bougierung die zirkuläre Schrumpfung der Transplantate begrenzt und eine Stenosierung sowie eine longitudinale Schrumpfung verhindert werden konnte. Die histologischen Befunde ergaben, daß frei transplantierte Jejunum-

mukosa/-submukosaschläuche von durchschnittlich 8 cm Länge bei Hunden und 5 cm bei den neugeborenen Kindern mit dem mediastinalen Wundgrund Granulationsgewebsrohre bilden, wobei die Innenauskleidung durch persistierendes Jejunumepithel, teils auch durch von den Ösophagusstümpfen her vorwachsendes Ösophagusepithel erfolgte.

Leider verstarben die beiden operierten Kinder ¼ bzw. 1 Jahr nach dem Eingriff an schweren Begleitfehlbildungen; dadurch war es aber möglich, auch die Ösophagusinterponate morphologisch aufzuarbeiten und die völlige Einheilung zu dokumentieren.

Vorteile dieser neuen Methode:

1) primär definitive Kontinuitätsüberbrückung,
2) geringere Problematik und geringeres Risiko gegenüber den Elongationsverfahren sowie den Ersatzplastiken,
3) möglicherweise kürzere Krankenhausverweildauer.

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The Outcome of Colonic Replacement of the Esophagus in Children

A. Ahmed[1] and L. Spitz[1]

A variety of methods have been devised in order to achieve a primary esophagoesophageal anastomosis in infants born with esophageal atresia. These include delayed primary anastomosis with (Howard and Myers 1965; Rehbein and Schweder 1971; Hendren and Hale 1975) or without (Cudmore 1978; Myers and Aberdeen 1979) the use of repeated bougienage to promote lengthening of the esophageal segments and the Livaditis (1973) esophageal myotomy. Nevertheless, there is a small group of patients in whom primary anastomosis cannot be achieved or in whom the esophagus has to be abandoned because of failure of the anastomotic technique. It is in this group of patients that replacement of the esophagus is required to restore continuity of the alimentary tract. In addition, esophageal replacement is occasionally necessary in children with intractable strictures secondary to gastroesophageal reflux or caustic esophageal injury.

Once the need for esophageal replacement has been established, a number of alternative methods are available. The aim of the replacement is to achieve continuity of the alimentary tract with a conduit which provides the best function with the fewest complications. None of the procedures currently performed is entirely free from complications in either the short or the long term. The advantages and disadvantages of the various types of replacements of the esophagus are shown in Table 1.

Although there are numerous reports available on the results of surgery for colonic replacement of the esophagus, few involve large numbers of cases in children and the follow-up is generally of short duration. We wish to report on an analysis of a large number of children undergoing colonic interposition followed up for a prolonged period of time.

Material and Methods

One hundred and twelve patients have undergone colonic replacement of the esophagus at The Hospital for Sick Children, Great Ormond Street, London, during the 30-year period 1952–1981. The majority (59) of the colonic interposition procedures were performed by Mr. D. Waterston, the remainder being carried out by a number of different surgeons during the period under review.

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Table 1. Advantages and disadvantages of various esophageal replacement methods

Type	Advantages	Disadvantages
Colonic interposition	Adequate length can usually be attained	Blood supply may be precarious (Postlethwait 1983); leaks/strictures from upper or lower anastomosis (25% on average) (German and Waterston 1976; Gross and Firestone 1967; Rodgers et al. 1978); redundancy of colon loop; kinks; slow transit time
Gastric-tube esophagoplasty	Good blood supply; adequate length; rapid transit of food	Long suture line; leaks from cervical anastomosis (50%) with strictures developing in 25% (Anderson and Randolph 1978; Cohen et al. 1974; Ein et al. 1973; O'Connor 1983)
Jejunal interposition	Size of intestine appropriate; good peristaltic activity	Vascular supply very precarious and adequate length difficult to attain (Dave et al. 1972)
Free jejunal graft	Size of intestine appropriate; good peristaltic activity (Ring et al. 1982)	Specialized technique for microvascular anastomosis required; prolonged operating time; high incidence of graft necrosis
Gastric interposition	Good blood supply; adequate length; ease of procedure	Bulk of stomach in mediastinum may cause respiratory problems (Sweet 1948); reflux and poor gastric emptying affect growth and development (Waterston 1969)

Indication

Esophageal atresia was the indication for the replacement in 92 children (59 male, 33 female), while extensive esophageal strictures were present in the remaining 20 children (15 male, 5 female). The precise indications for esophageal replacement are listed in Table 2. Esophageal atresia was classified as follows:

Type A – atresia with distal tracheoesophageal fistula
Type B – atresia with proximal tracheoesophageal fistula
Type C – atresia with proximal and distal fistulae
Type D – isolated atresia without fistula

Patients in the stricture group had intractable peptic, caustic, or congenital esophageal strictures.

Age

The age distribution of the patients at the time of the colonic replacement is shown in Fig. 1. In 85 children the procedure was performed within the first 2

Table 2. Indications for colonic interposition

	Esophageal atresia			
	Type A	Type B	Type C	Type D
Large gap	33	2	2	28
Failed primary anastomosis	17	1	2	1
Failed delayed primary anastomosis	5	1	–	–
	55	4	4	29

	Esophageal stricture		
	Reflux stricture	Caustic	Congenital stenosis/ stricture
Failure of antireflux procedure and repeated esophageal dilatations	6	–	1
Failed repeated esophageal dilatations	3	3	–
Intractable stricture	5	1	–
Recurrent stricture after excision and esophageal dilatations	–	–	1
	14	4	2

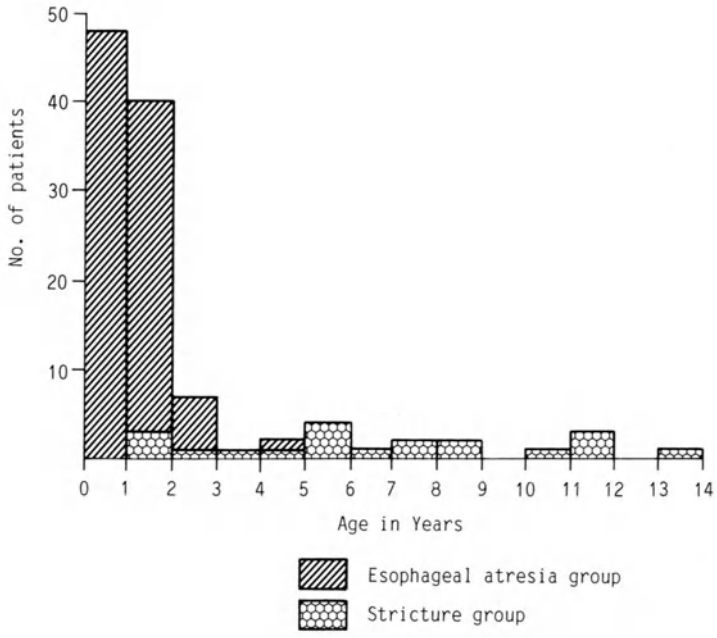

Fig. 1. Age distribution of patients according to diagnosis at the time of colonic interposition

years of life; these included two infants undergoing primary colonic interposition without preliminary cervical esophagostomy before 2 months of age. Colonic replacement for esophageal stricture was never performed during the first year of life, the cases being evenly distributed between 1 and 14 years of age.

Previous Surgery

Of the 92 patients with esophageal atresia, 90 had previously undergone a left cervical esophagostomy and a feeding gastrostomy. The two infants who underwent primary colonic replacement had had a feeding gastrostomy only. Three patients in the stricture group had a cervical esophagostomy and two a feeding gastrostomy performed prior to the replacement procedure.

Type of Procedure

Eighty-two of the interpositions involved transthoracic replacement of the esophagus (Waterston type) with the proximal anastomosis to the cervical esophagus; the distal anastomosis was to the blind esophageal stump in 73 patients and to the stomach in nine. A partial intrathoracic replacement was employed in 18 children, with the proximal anastomosis within the left thoracic cavity and the distal anastomosis to the distal esophageal stump in two cases and to the stomach in 16. The remaining ten procedures consisted of a retrosternal placement of the colonic interposition. Two colon loops had to be abandoned prior to reconstruction due to irreversible ischemic damage. The transverse colon was used in 88 patients, the left colon in 19, and the right colon in four. Operative details were inadequate in one case. All colon loops were placed in the isoperistaltic direction.

Associated Anomalies

Associated congenital anomalies were identified in 35 of the 92 children with esophageal atresia (38%). A total of 68 anomalies were encountered in the 35 patients as shown in Table 3.

Table 3. Incidence of associated anomalies in esophageal atresia group

1. *Gastrointestinal anomalies* – 20	
Imperforate anus:	
high type	6
low type	5
Duodenal atresia	5
Congenital pyloric stenosis	1

Table 3 (continued)

Annular pancreas	2
Meckel's diverticulum	1
2. *Musculoskeletal anomalies* – 16	
Vertebral	3
Radial	2
Absent proximal humerus	1
Madelung's deformity	1
Wrist deformity	1
CDH	1
Thumb	5
Polydactyly	1
Rib	1
3. *Cardiovascular anomalies* – 6	
PDA	1
Fallot's	1
VSD	3
Coarctation of aorta	1
4. *Craniofacial anomalies* – 3	
Choanal atresia	2
Ptosis	1
5. *Pulmonary thorax anomalies* – 7	
Cleft larynx	2
Subglottic stenosis	1
Agenesis right lung	1
Single-lobed right lung	1
Pulmonary lymphangiectesis	1
Congenital emphysema	1
6. *Genitourinary anomalies* – 10	
Renal agenesis (L)	2
Dysplastic kidney	1
Cystic, nonfunctioning kidney	1
Hydronephrosis	2
Reflux	1
Bladder neck obstruction	1
Hypospadias	2
7. *Central nervous system anomalies* – 2	
Hydrocephalus	2
8. *Chromosomal anomalies* – 3	
Down's syndrome	3
9. *Miscellaneous* – 1	
Hypothyroidism	1

Results

Mortality

There were 15 deaths (13.4%), all in the atresia group (Fig. 2). Nine deaths occurred early in the postoperative period: three were directly due to necrosis of the interposition, four were secondary to anastomotic leakage or disruption, and two were the result of respiratory failure. Three deaths followed secondary replacement for "failed" colonic interpositions. Two of these patients died early in the postoperative period, while the other died 4 years after a gastric-tube esophagoplasty from hematemesis secondary to gastric erosions. The one unrelated death was due to acute pyelonephritis in an infant with a high anorectal agenesis and a rectourethral fistula. There were two late deaths – one in a severely retarded child, 2 years after a successful interposition, from respiratory failure, hydrocephalus, and tetralogy of Fallot, and the other, 11 years later, from respiratory failure secondary to agenesis of the right lung and severe kyphoscoliosis.

Failure of Colonic Interposition

Failure of the colonic interposition was encountered in 16 patients (14.3%) and accounted for six of the deaths (Fig. 3). Necrosis of the interposition secondary to vascular insufficiency was the cause of failure in nine patients and resulted in three deaths. In the remaining six patients, the colon was excised and a subsequent secondary replacement was performed. Two of these patients died. Five patients, one of whom died, developed intractable strictures requiring secondary interpositions. In two patients the interpositions failed to function, necessitating further reconstruction.

Anastomotic Complications

Leakage of the proximal esophagocolonic anastomosis was the most common complication, developing in 54 patients (48.2%). Five of these anastomoses had to be totally reconstructed, while a further three required surgical closure. The remaining fistulae closed with conservative management. Strictures developed at the site of the anastomosis in 23 of these patients and were also encountered in a further 11 patients whose anastomoses did not leak clinically. A total of 34 patients (30.3%), therefore, developed a stricture at the proximal esophagocolonic anastomosis.

The incidence of leakage at the cologastric or distal coloesophageal anastomosis was much less frequent, encountered in 16 patients (14.3%). In four patients the leakage was directly responsible for death as a result of empyema and pneumonia. With the exception of a single patient who required revision of the anastomosis, all the remaining fistulae closed with conservative treatment. Eleven patients

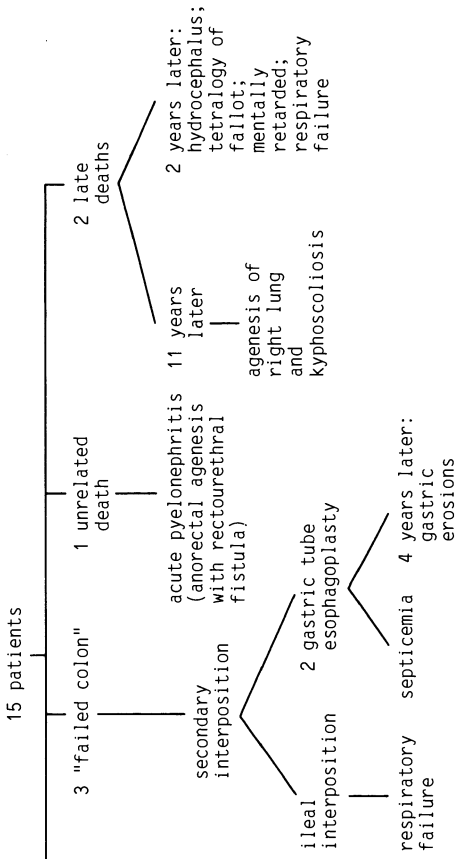

Fig. 2. Analysis of deaths of 15 patients

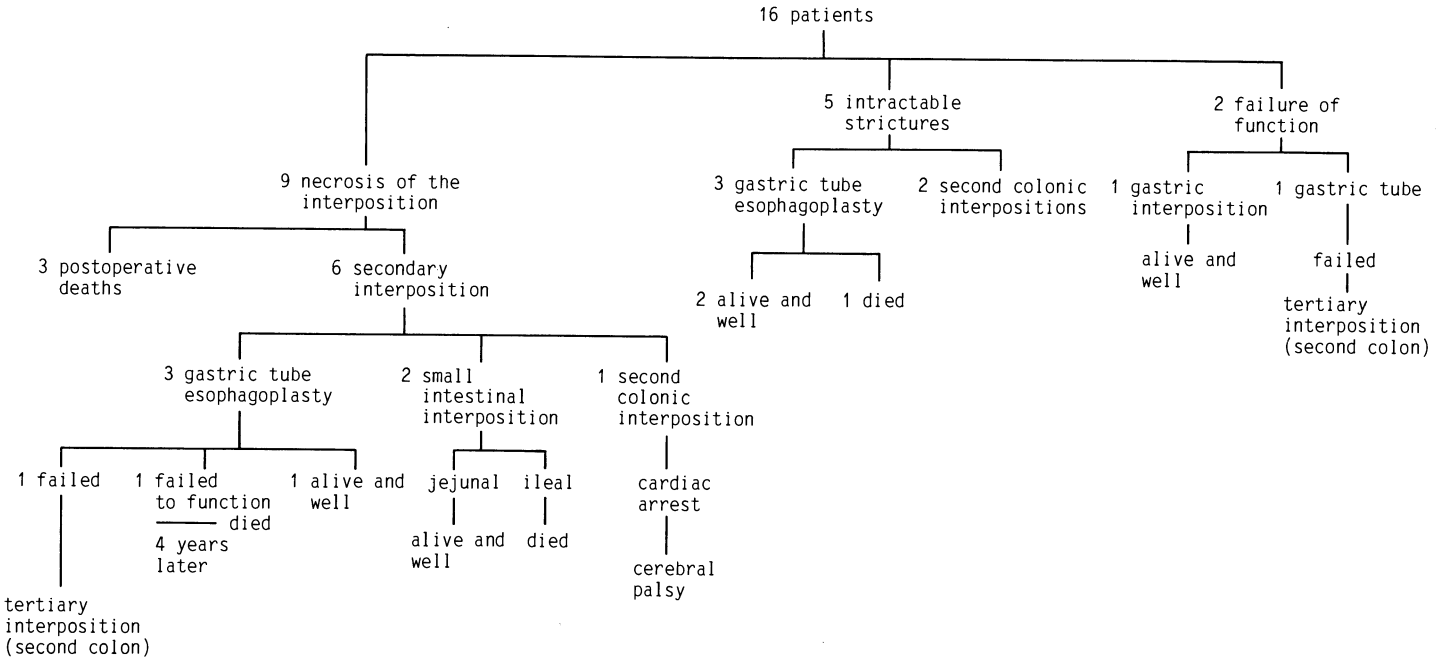

Fig. 3. Reasons for failure of colonic interposition in 16 patients

Table 4. Anastomotic complications related to type of interposition and etiology of the condition

Site of anastomotic complication	Total	Percent	Transthoracic interposition (Waterston type) (82)		Retrosternal interposition (10)		Intrathoracic partial replacement (18)	
			EA – 79	Stricture – 3	EA – 9	Stricture – 1	EA – 2	Stricture – 16
Leakage								
Proximal	54	48.2	41	2	7	1	1	2
Distal	16	14.3	12	1	–	–	–	3
Stricture								
Proximal	34	30.3	26	1	3	–	1	3
Distal	11	9.8	6	–	2	–	1	2

EA = Esophageal atresia

(9.8%) developed strictures at the distal anastomosis, two directly related to anastomotic leakage.

The incidence of anastomotic complications related to the type of procedure and the primary pathology is shown in Table 4, while the incidence of complications related to staging of the interposition is shown in Table 5. The frequency of anastomotic leakage or stricture formation could not be related to single- or multiple-stage procedures.

Revision of the proximal esophagocolonic anastomosis was required in 20 patients, five of whom had to undergo secondary revisions. The distal anastomosis required primary revision in six patients and secondary revision in one. Postvagotomy syndrome developed in one child. Diarrhea was a significant problem for four patients, while malabsorption due to blind-loop syndrome was present in one.

Long-term Graft-related Complications

Mild colitis detected either by radiological examination or at endoscopy was documented in five patients, two of whom had iron-deficiency anemia. All cases resolved with conservative therapy, including one patient who required a pyloroplasty for gastric stasis. Redundancy of a sufficient degree to warrant resection was observed in six cases. A significant esophageal pouch developed in six patients, three of whom required revision surgery. Kinking of the colonic interposition was generally associated with other problems, i.e., diaphragmatic hernia, redundancy, and esophageal diverticulum.

Table 5. Staging of procedure and incidence of anastomotic leakage, stricture, failure of interposition, and death

	No. of cases	Anastomotic leak		Anastomotic stricture		Failure of interposition			Death		
		Prox.	Dist.	Prox.	Dist.	Necrosis	Stricture	Failed to function	Post-op.	Late related	Not related
1. Single stage Isolation of colon and proximal and distal anastomosis	70	32	13	20	9	2	4	2	7	1	2
2. Two-stage A a) Isolation of colon, distal anastomosis and neck colostomy b) Proximal anastomosis	28	15	3	9	2	5	1	0	2	1	1
3. Two-stage B a) Isolation of colon with abdominal colostomy b) Proximal and distal anastomosis	8	1	0	1	0	2	0	0	0	1	0
4. Three stages a) Isolation of colon and abdominal colostomy b) Distal anastomosis and neck colostomy c) Proximal anastomosis	5	5	0	4	0	0	0	0	0	0	0
5. Insufficient details	1	1									
	112	54	16	34	11	9	5	2	9	3	3

Outcome

Eighty-seven patients had a successful colonic interposition (excluded are the deaths and secondary replacements involving other parts of the gastrointestinal tract). Ten patients could not be traced for follow-up. The follow-up period for the remaining 77 patients ranged from 3 to 24 years (see Table 6). The results were classified as follows:

Excellent: the patients were entirely asymptomatic and could accept a normal diet (43 patients)

Good: the patients were asymptomatic for most of the time but suffered occasional minor disturbances with dysphagia (27 patients)

Fair: the patients suffered significant dysphagia or required frequent dilatations (7 patients)

The weight and height of the surviving patients is shown in Table 7.

Table 6. Outcome for 77 patients undergoing successful colonic interposition

Follow-up period (years)		Excellent	Good	Fair
3– 5		4	4	2
5–10		16	7	2
10–15		11	7	2
15–20		6	5	0
>20		6	4	1
	Total	43	27	7
Diagnosis				
Esophageal atresia		34	24	6
Esophageal stricture		9	3	1

Table 7. Weight and height (long-term follow-up)

Percentile	Weight (no. of patients)	Height (no. of patients)
<3°	21	13
3–10°	21	9
10–25°	9	11
25–50°	15	15
50–75°	4	18
75° +	5	7

Discussion

Bircher (1907) described the first attempts at bridging the gap between the obstructed esophagus and the stomach by the construction of skin-lined tubes. Subsequently, the entire stomach (Adams and Phemister 1938; Sweet 1945; Garlock 1948) and tubes fashioned from the stomach (Beck and Carrell 1905; Jianu 1912; Gavriliu and Georgescue 1951; Heimlich 1957; Sanders 1962; Burrington and Stephens 1968; Anderson and Randolph 1978; Cohen et al. 1974), the small bowel (Roux 1907; Yudin 1944; Longmire 1951; Merendino and Thomas 1958; Grimes 1967a), and the colon (Kelling 1911; Lundblad 1921; Sandblom 1948; Sherman and Waterston 1957; Waterston 1969; Grimes 1967b) have been used.

The use of a colonic segment has become the most widely accepted method of total or partial replacement of the esophagus in children. The colon has proven to be relatively acid resistant, and significant ulceration in the interposed colon is unusual. Many operations have been devised for placing various segments of the colon intrapleurally (Sandblom 1948; Waterston 1969), retrosternally (Javid 1954; Dale and Sherman 1955; Gross and Firestone 1967; Schiller et al. 1971), subcutaneously (Lundblad 1921), and in the normal esophageal route in the posterior mediastinum (Belsey 1965; Akiyama et al. 1975; Freeman and Cass 1982). The two most widely used anatomical routes are the left intrathoracic and the retrosternal. The procedure involved carries a significant mortality, and the morbidity

Table 8. Complication rates from colonic interposition from various series reported in the literature

Author(s)	Year	No. of patients	Mortality (%)	Ischemia of graft (%)	Anastomotic leakage (%)	Stricture (%)
De Boer	1964	14	14.3	–	28.6	28.6
Gross and Firestone	1967	47	8.5	4.2	12.8	14.9
Othersen and Clatworthy	1967	11	–	–	36.3	27.3
Sieber and Sieber	1968	10	–	–	–	50.0
Schiller et al.	1971	29	–	3.4	34.5	24.0
Azar et al.	1971	60	8.0	3.3	25.0	30.0
Singh and Rickham	1971	12	8.3	8.3	41.6	8.3
Soave	1972	32	15.6	3.1	56.2	N/A
Martin	1972	21	9.5	–	19.0	28.5
Rodgers et al.	1978	13	–	–	38.5	23.0
Freeman and Cass	1982	16	12.5	–	12.5	12.5
Lindahl et al.	1983	19	15.8	21.0	47.3	21.0
Present series	1984	112	13.4	8.0	48.2	30.3
Collective review:						
Postlethwait	1983	2067	11.4	8.0	25.0	10.0

from ischemic necrosis, anastomotic leakage, and stricture formation is not inconsiderable. The statistics on these complications in children from a number of the larger series reported in the literature are shown in Table 8.

The mortality associated with colonic replacement in children over the age of 6 months is low. Most of the reported deaths occurred in small infants, where regurgitation and aspiration are of much greater potential hazard than in older children or in adults. In our series 12 (10.7%) of the 15 deaths were directly or indirectly related to the operative procedure. Half of these infants had major associated congenital anomalies. The three unrelated deaths also suffered from severe associated anomalies.

There were nine instances of ischemic necrosis of the interposed colon, an 8% incidence. Three of the deaths were directly caused by this complication. Early diagnosis and prompt treatment by excision of the ischemic graft can prevent fatalities.

The overall incidence of proximal anastomotic leaks was high (48%). The incidence of leakage varied according to the technique employed. The leak rate for retrosternal interposition was 80% (eight of ten cases); for transthoracic, 52.4% (43 of 82 cases); and for intrathoracic procedures, 16.6% (three of 18 cases). The high incidence of cervical anastomotic problems in colonic interposition procedures may be ascribed to the fact that the anastomosis is performed farthest from the blood supply and that it is easily angulated and placed under tension. Some authors recommend resection of part of the manubrium or the head of the clavicle and division of the sternohyoid, sternothyroid, and sternal head of the sternocleidomastoid muscle (Gross and Firestone 1967; Martin and Flege 1964; Rodgers et al. 1978) in the retrosternal procedure to allow for more room at the thoracic inlet and to minimize any pressure on the esophagocolonic anastomosis. Compression at the diaphragm should be avoided, and redundancy of the colonic loop is undesirable. Staging of the replacement procedure in various ways (Table 3) does not appear to be an effective means of reducing the incidence of leaks or subsequent stricture formation. It may, however, be useful if the vascularity of the colon segment is in any way doubtful (Sherman and Waterston 1957; Martin and Flege 1964).

Strictures of the proximal esophagocolonic anastomosis developed in 42% of cases following leaks, but they were also encountered in patients without a clinically diagnosed leakage. The incidence of stricture formation was unrelated to the method of interposition. Three-quarters of the strictures were treated by operative revision of the anastomoses, while five patients ultimately required a secondary interposition for intractable strictures. Endoscopic dilatations were notably unsuccessful and carried the additional risk of perforation (Gross and Firestone 1967). Since the advent of fiberoptic endoscopes this hazard has been virtually eliminated.

Trauma to the recurrent laryngeal nerves is a potentially lethal complication but it is most frequently due to neuropraxia and is recoverable. This complication was encountered in one of our cases. It was responsible for two deaths in Freeman's series (Freeman and Cass 1982). We would advocate deliberate iden-

tification and careful preservation of the recurrent laryngeal nerve prior to any mobilization of the cervical esophagus.

Motor activity of the transposed colon remains a subject of controversy. Early investigators, using fluoroscopy, cineradiography, and manometry, failed to demonstrate the presence of motor activity in the colon graft, and it was assumed that the transplant acted as a passive conduit emptying by gravity (Othersen and Clatworthy 1967; Sieber and Sieber 1968; Martin and Flege 1964). There appeared to be no difference in function between the colon placed in the isoperistaltic or antiperistaltic position (Sieber and Sieber 1968). More recent studies report evidence of sequential or propulsive waves in the interposed segment, promoting the evacuation of content from the colon into the stomach and clearing refluxed gastric content from the colon (Jones et al. 1973; Miller et al. 1975; Clark et al. 1976; Benages et al. 1981). These observations are important and should encourage surgeons to interpose the transplanted colon in the isoperistaltic direction.

Follow-up studies for periods ranging from 3 to 24 years confirm the findings of other major series that indicate the functional adequacy of the interposed colon. A satisfactory result was attained in 70 of the 77 patients responding to the questionnaire, i.e., 90% of responders and 68.6% overall (excluding patients lost to follow-up).

We have observed that many of the initial problems associated with colonic reconstruction, such as respiratory infections and swallowing difficulties, are temporary phenomena which improve with increasing length of follow-up. It is, however, possible to predict with a fair degree of accuracy at the 3-year follow-up evaluation the final outcome of the procedure.

It has also been noted that the results of colonic replacement for strictures are better than those for esophageal atresia. This may be partially explained by the fact that a large proportion of cases (38%) with atresia had associated congenital anomalies which contributed to the mortality and morbidity with the various surgical procedures. Moreover, the inherent motility disturbances in the esophagus in atresia cases are also responsible for some of the respiratory and swallowing problems.

Despite the overall satisfactory results in our series, the colon transplant patients tended to be small and underweight (68% below the 25th percentile of weight, 45% below the 25th percentile of height). German and Waterston (1976) evaluated two similar groups of patients with parallel anatomical problems, esophageal replacement and repaired esophageal atresia. The comparison between the two groups showed no significant difference in weight gain but they could not demonstrate a catch-up phase as reported (Louhimo et al. 1969). Longitudinal weight gain of both groups tended to remain between the 3rd and 25th percentiles over the entire 12-year period of their study. They further observed that the colon transplant group did have a consistently lower birth weight than the esophageal atresia group, which may indicate that these patients start out life small-for-date.

The high mortality and morbidity associated with colonic interposition in infancy and childhood have prompted us to evaluate total gastric transposition as

an alternative method of esophageal replacement. The early results of transhiatal gastric interposition in infants with esophageal atresia have been reported (Spitz 1984) and are encouraging.

Summary

An analysis of 112 children undergoing colonic replacement of the esophagus over a 30-year period is presented. The indication for esophageal replacement was atresia in 92 children and intractable stricture (peptic, caustic, or congenital) in 20. The procedure consisted of a transthoracic replacement of the entire esophagus in 82 cases and a partial replacement in 18, while a retrosternal replacement was used in ten cases. Two colon loops had to be abandoned prior to reconstruction due to irreversible ischemic damage. There were 15 deaths (13.4%) – all in the atresia group. Failure of the colonic graft was encountered in 16 patients (14.3%) and accounted for six of the deaths. Leakage of the proximal esophagocolonic anastomosis occurred in 54 cases (48.2%). Strictures of this anastomosis developed in 34 cases (30.3%). A total of 20 patients required operative revision of the anastomosis. The final outcome was excellent in 43 of 77 cases followed up for up to 24 years postoperatively (55.9%), good in 27 cases (35%), and only fair in seven cases (9.1%).

Résumé

Les auteurs présentent l'analyse de 112 observations d'enfants ayant subi une œsophagoplastie colique sur une période de 30 ans.

Les indications pour cette œsophagoplastie ont été l'atrésie dans 92 cas et un rétrécissement impossible à traiter (d'origine peptique, caustique ou congénitale) chez 20 enfants. La technique a consisté en un remplacement trans-thoracique de tout l'œsophage dans 82 cas et un remplacement partiel dans 18 cas; un remplacement rétrosternal a été effectué dans 10 cas. Il y a eu 15 décès (13,4%), tous dans le groupe des atrésies. L'échec du transplant colique a été recontré chez 16 patients (14,3%) et responsable de 6 des décès. Le lâchage de l'anastomose œsophago-colique proximale est survenu dans 54 cas (48,2%). Des rétrécissements de cette anastomose se sont développés dans 34 cas (30,3%). Un total de 20 patients ont nécessité une révision chirurgicale de l'anastomose.

Le résultat final a été excellent chez 43 sur 77 patients suivis plus de 24 ans après l'opération (55,9%), bon chez 27 patients (35%) et seulement médiocre dans 7 cas (9,1%).

Zusammenfassung

Die über 30 Jahre gesammelten Erfahrungen mit 112 Kindern, die eine Kolonersatzplastik des Ösophagus erhalten hatten, werden analysiert. Die Indikation

für einen Ösophagusersatz erfolgte in 92 Fällen aufgrund einer Ösophagusatresie und in 20 Fällen aufgrund einer nicht behandelbaren kaustischen oder peptischen Striktur. Ein transthorakaler Ösophagusersatz wurde in 82 Fällen, ein retrosternaler in 10 Fällen vorgenommen. 15 Todesfälle (13,4%) betrafen die Gruppe der Ösophagusatresie; 16mal wurden Nekrosen des Koloninterponats festgestellt (14,3%), 6 dieser Fälle gehörten zu denen mit letalem Ausgang. Eine Dehiszenz der proximalen ösophagokolischen Anastomose zeigte sich bei 54 Patienten (48,2%). Strikturen dieser Anastomose entwickelten sich in 34 Fällen (3,3%). Insgesamt war bei 20 Patienten eine operative Revision der Anastomose notwendig. Das Endergebnis war bei 43 von 77 Patienten, die über 24 Jahre postoperativ nachuntersucht wurden, ausgezeichnet (insgesamt 55,9%). Ein gutes Ergebnis gab es bei 27 Patienten (35%), nur mäßig war das Resultat in 7 Fällen (9,1%).

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Gastric Tube Esophagoplasty

K. D. ANDERSON[1]

History

The esophagus was a surgical no-man's land until Czerny first successfully resected its cervical portion in 1877. Since that time many and varied conduits (Kelling 1911; Merendino and Dillard 1955; Sweet 1945) have been used to replace the esophagus destroyed by stricture or malignancy. Typically, advances in the surgical care of children have lagged behind advances for adults. Infants with long-gap esophageal atresia are no exception. The rare survivors of this anomaly underwent multiple procedures to fashion skin-lined tubes of skin placed beneath the skin of the anterior thorax (Gross 1953). Although these children were able to swallow food they were forced to massage it down to the stomach and they were required to live with their hideous appearance. In 1955, Dale and Sherman described the use of a portion of the colon placed substernally in a child with esophageal atresia, and the modern era of esophageal substitution for children began. The colon is still the most widely used substitute for the esophagus (Campbell et al. 1982; Gross and Firestone 1967; Neville and Nahem 1983; Waterston 1964). However, the numerous associated problems led to a search for a better substitute. Following the pioneering work of Gavriliu (see for example Gavriliu 1975 and Heimlich (1966), Burrington and Stephens (1968) and later Anderson and Randolph (1973) and Cohen et al. (1974) popularized the use of the reversed gastric tube for esophagoplasty in the child. In the author's institution, a total of 42 patients have required esophageal replacement since 1965. Nineteen of these patients underwent colon transposition and 24 received reversed gastric tube. Fourteen of the latter patients underwent esophagoplasty because of long-gap esophageal atresia, and of these ten were available for follow-up for up to 14 years.

Indications

There are four classic indications for esophageal substitution. The first is esophageal atresia with or without tracheoesophageal atresia with a long gap between the upper and the lower pouch. As experience is gained with secondary anastomosis following a period of 6–8 weeks of growth, during which the disparate esophageal

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pouches may approach each other (Woolley 1980), cervical esophagostomy and later esophageal replacement is becoming less common. The technique of circular myotomy (Livaditis et al. 1972; Ricketts et al. 1981) has been used successfully to increase the length of the upper pouch. In spite of considerable progress in these techniques, they cannot be universally applied, and replacement will still occasionally be required.

Intractable strictures secondary to ingestion of caustic alkali or acids are an all too frequent occurrence in the United States, where caustic materials are attractively packaged and available in a plethora of brands. A method of stenting the esophagus has been described (Hill et al. 1976) to avoid the necessity for esophageal replacement. However, for injury over more than a few centimeters it is doubtful if full re-epithelialization can occur, and replacement is still required if dilatation is unsuccessful.

An older indication for esophageal replacement was the presence of esophageal varices secondary to portal hypertension. New techniques of portosystemic shunts (Altman 1976; Clatworthy and Boles 1959) and endosclerosis (Lilly et al. 1982) have eliminated the need for removal and replacement of the esophagus.

A rare indication for esophageal replacement remains (Skinner and De Meester 1976); strictures secondary to gastroesophageal reflux rarely occur since chalasia is now almost universally recognized and treated early. However, the occasional case of a neglected stricture will require esophageal replacement.

Timing of Surgery

With the increasing skills of anesthesiologists and nursing personnel and the refinement of surgical techniques, almost any procedure can be performed in the neonate. However, the small stomach of the neonate with pure esophageal atresia precludes the creation of a gastric tube in this early age group. Gastrostoma feedings can support the nutritional needs of the infant and enlarge the stomach. As long as sham feedings are given by mouth, the infant should have little difficulty in learning to feed. There is therefore no urgency to perform the procedure in early infancy. We elect a weight of around 10 kg as the "ideal" weight at which to perform the major surgery.

Is the Gastric Tube the "Ideal" Substitute?

The "ideal" substitute must have the following characteristics:

1. The operative procedure should be relatively simple technically and adaptable to the child's anatomy.
2. The esophageal substitute should act as an efficient conduit from mouth to stomach.
3. Ventilation must not be impaired in the small child, so the substitute itself must be tubular and must remain so long-term.

4. Reflux of gastric contents into the substitute esophagus must be minimal or correctable, or the mucosa itself must be resistant to corrosion by gastric juice.
5. The conduit should grow with the child so that nutritional needs can be satisfied into adulthood without the need for additional surgical procedures.

Both colon and gastric-tube substitutes act as good esophageal conduits, though effective peristalsis does not occur in either organ. The operative technique is somewhat simpler for creation of the gastric tube, and the blood supply of the stomach is superior to that of the colon. The colon tends towards redundancy as the child grows; this is often an incidental observation, however, and not necessarily associated with symptoms. Since a longer length of colon can be left in an intra-abdominal position, reflux is somewhat less a problem than with the gastric tube. However, the colon mucosa, unlike the gastric mucosa, has a very inefficient barrier to acid. Reflux, therefore, when it occurs into the colon segment, may have dire consequences, and ulceration occurs much more commonly in the colon than in the gastric tube (Anderson et al. 1975; Malcom 1968; Singh and Rickham 1977).

Technique of Gastric Tube Esophagoplasty

Selection of the site for placement of the feeding gastrostoma is important. For ease of construction of the gastric tube the gastrostoma is placed well away from the greater curvature. This may be somewhat difficult in the contracted stomach of the neonate with pure esophageal atresia, but the opening can be made immediately adjacent to the lesser curvature in mid stomach without untoward effect. The cervical esophagostoma is placed in the left neck.

The operative field includes the left side of the neck, the thorax including the entire sternal area, the left arm, and the entire abdomen. Satisfactory exposure is achieved by a transverse upper abdominal incision, dividing both rectus muscles. The gastrostoma is taken down from the abdominal wall and sutured closed. (It will be used again at the conclusion of the procedure.) The gastrocolic omentum is divided away from the gastroepiploic arcade. The right gastroepiploic artery is divided, and a stab wound is made in the anterior and posterior gastric walls at this point (Fig. 1A). A chest tube of appropriate size (18–24 French, depending on the infant's size) is inserted through this stab wound and held in position by a stay suture in the distal stomach, tied around the tube. Clamps to hold the chest tube in position are not recommended. Using the GIA stapler (American Surgical Corp.) the anterior and posterior walls of the stomach are stapled together in two rows with an incision between to create the first part of the tube (Fig. 1B). Blood loss is minimal. It is important for the assistant to hold the greater curvature gently as this is done, since the gastroepiploic arcade tends to slew forward into the jaws of the stapler. Three to four cuts are made with the stapler, parallel to the greater curvature, taking care not to advance so far at the upper end as to narrow the channel between the tube and the rest of the stomach. Short gastric vessels and the spleen are left intact. The staple lines on both tube and stomach are reinforced with nonabsorbable 4–0 suture (Fig. 1C).

The gastric tube can be placed in a retrosternal or retrohilar position. If the substernal route is elected, a thoracotomy is not performed and a suprasternal incision is used. The substernal tunnel is made by blunt dissection from above and below. The author prefers the transthoracic route through the 6th intercostal space. Once the chest is open a cervical incision is made, extending posteriorly from the cervical esophagostomy but not dissecting it free at this point. Finger dissection in the neck facilitates exposure of Sibson's fascia in the thoracic inlet. Working from cervical and thoracic incisions, the operator's fingers explore the position of the subclavian vessels, and an opening is made in the fascia either anteriorly or posterior to these vessels, depending on

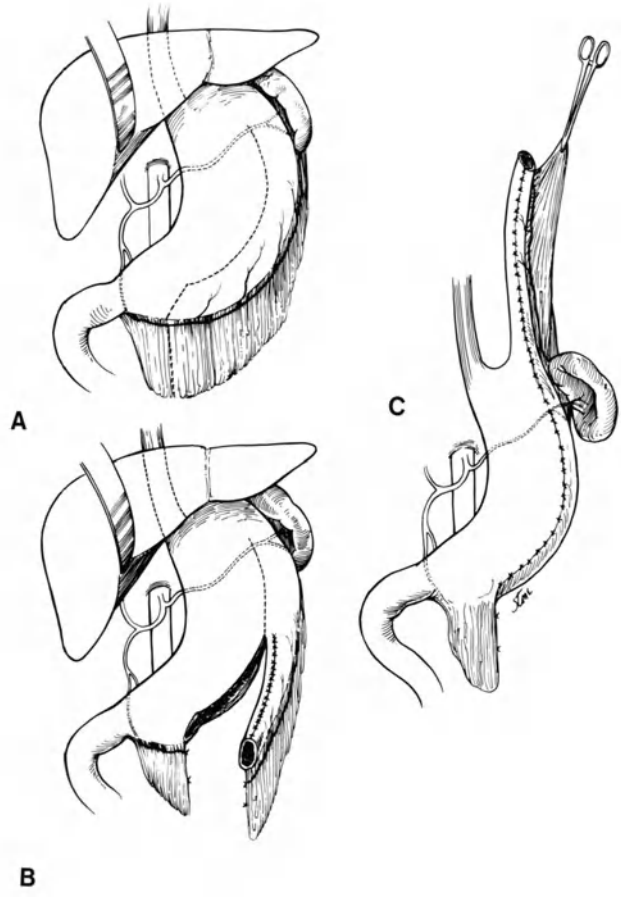

Fig. 1. A Outline of gastric tube along greater curvature of stomach. **B** Partially made tube; stapler is used to cut and anastomose tube simultaneously. **C** Completed tube, reinforced with nonabsorbable sutures, can be placed substernally or in the retrohilar area

which space is larger. The opening is dilated bluntly. An incision is made in the diaphragm, anterolateral to the aorta, and dilated just enough to accommodate the gastric tube and its vascular arcade without constriction. The tube is drawn into the chest, gently placed behind the lung, and drawn into the neck. The pedicle remains in a constant position to avoid twisting or kinking of the vessels. Any extra length is left in the abdomen to decrease reflux. Two or three nonabsorbable sutures are placed between serosa and diaphragm to fix the tube in position. No sutures are placed at the apex of the chest. At this point a decision is made to complete the procedure in one stage or to perform the tube-esophageal anastomosis at a later date. Use of the stapler has reduced the time taken, and the entire procedure can usually be completed in one stage. The cervical esophagostoma is taken down at this point and approximately 2–3 cm of esophagus are dissected from surrounding tissue. A single layer of 4–0 nonabsorbable sutures is used for the cervical anastomosis. The gastrostoma is replaced in the old site and all incisions are closed. The neck is drained with a Penrose drain and a chest tube drains the thorax. If the left gastroepiploic artery is not suitable for the vascular pedicle, the tube can be fashioned in the reverse direction based on the right gastroepiploic vessels.

Broad spectrum antibiotic coverage continues for 72 h and peripheral parenteral nutrition is given until gastrostoma feedings are started, usually on the 5th day. Barium swallow is performed on the 7th postoperative day before removal of the chest tube. Once integrity of the tube has been demonstrated, oral feedings are begun. Small, frequent gastrostoma feedings are required

initially until the stomach returns to a normal size. In cases of cervical anastomotic leak, oral feeding is delayed. In most instances these leaks close spontaneously within a week or two.

Results and Follow-up

Ten of our 14 patients who had gastric-tube esophagoplasty for esophageal atresia were available for follow-up to 14 years. Two patients were lost to follow-up after 2–3 years but were doing well when last seen. Two patients died: one patient, who had been born at 28 weeks gestation, died at 2 years of age of chronic lung disease; the other had an unrecognized perforation of the thoracic portion of the gastric tube. Indications for dilatation and the circumstances surrounding this patient's death are unknown.

Eight children are growing and developing normally. Two children are retarded (one has trisomy 21) but their nutrition is satisfactory. Few of the children exceed the 25th percentile for weight. Minimal swallowing difficulties are reported and all children eat a full meal with the rest of the family. Fibrous meats need to be chewed thoroughly but no patient avoids these foods. One patient required a single late dilatation as he entered his adolescent growth spurt, but in general dilatation of the upper anastomosis has not been required beyond the first 8–12 months postoperatively.

Complications such as salivary fistulas and mild anastomotic strictures resolved spontaneously, and only patients operated on for lye stricture needed any secondary anastomotic revision. One patient had a persistent tiny leak which did not interfere with oral feedings; this was closed surgically after 1 year. A single patient had perforation of the gastric tube during esophagoscopy in our institution; this was recognized and closed surgically without further problems. One patient had an ulcer in a redundant portion of the tube in the chest (Anderson et al. 1975). This healed with antacid therapy and the patient has been well for 10 years. Two patients require propping up at night to avoid nocturnal coughing secondary to reflux. Restricting fluids 2 h before bedtime virtually eliminates this problem and no patients have developed pneumonia. Reflux is a decreasing problem as the children get older.

The gastric tube has maintained its tubular form in all cases studied by barium swallow, and emptying occurs promptly. The stomach resumes its normal size within 6 months of surgery, and by this time regular-sized meals can be enjoyed by all the children.

Conclusions

The gastric tube serves as a highly satisfactory esophageal substitute for the growing child. Complications are early, usually technical in nature, and fairly easily handled in most instances. Moderate follow-up shows a decrease in problems secondary to reflux. Long-term follow-up will be needed to define late complications before the gastric tube can be claimed to be truly the ideal esophageal substitute.

Summary

After a short review of the historical development of replacement of the esophagus, the possibilities, techniques, and results of gastric-tube esophagoplasty are discussed. Ten of 14 patients were followed up for 14 years, two were lost to follow-up and two died. Fistulas, anastomotic stictures, and also reflux could be treated successfully. A single perforation by endoscopy was surgically corrected. A peptic ulcer in the gastric tube was healed with antacid therapy. The gastric tube can be assumed to be a sufficient esophageal substitute. Long-term follow-up is still necessary.

Résumé

Après une courte revue concernant l'évolution historique des œsophagoplasties, les possibilités techniques et les résultats des remplacements œsophagiens par tube gastrique sont discutés. Dix sur 14 patients ont été suivis plus de 14 ans, 2 ont été perdus de vue et 2 sont morts. Les fistules, les rétrécissements anastomotiques et aussi le reflux ont pu être traités avec succès. Une perforation unique lors d'une endoscopie a pu être traitée chirurgicalement. Un ulcère peptique dans le tube gastrique a guéri par une thérapeutique anti-acide. Le tube gastrique peut être considéré comme un substitut œsophagien convenable. Une étude à long terme est néanmoins nécessaire.

Zusammenfassung

Nach einem kurzen historischen Überblick über die Entwicklung des Ösophagusersatzes werden die Möglichkeiten, Techniken und Ergebnisse der Magenschlauchösophagoplastik diskutiert. 10 von 14 Patienten wurden über 14 Jahre nachuntersucht; 2 konnten nicht nachuntersucht werden und 2 starben. Fisteln, Anastomosenstrikturen und ebenso ein gastroösophagealer Reflux konnten erfolgreich behandelt werden, eine einzelne Perforation bei einer Endoskopie wurde chirurgisch korrigiert, ein peptisches Ulkus im Magenschlauch heilte durch antazide Therapie. Der Magenschlauch kann als genügender Ösophagusersatz angesehen werden. Langzeitnachuntersuchungen sind allerdings noch notwendig.

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The Significance of Tracheal Stenosis in Esophageal Atresia

P. WURNIG[1], K. MANTEL[2], and W. CH. HECKER[2]

It has long been known that children with esophageal atresia may suffer from respiratory problems for years (Daum 1970). Studies revealed that esophageal atresia is frequently associated with tracheal malacia (Benjamin et al. 1976), with anatomical disturbances of the tracheal skeleton and the tracheal mucosa (Emery and Haddadin 1971). It was suspected that the large upper pouch impeded tracheal growth within the narrow upper thoracic aperture (Davies and Cywes 1978), but no therapeutic conclusions were drawn, although life-threatening apneic and hypoxic spells had been observed. Several investigators found that a considerable number of patients operated on for esophageal atresia had external tracheal obstruction by the innominate artery (Berdon et al. 1969; Fearon and Shortreed 1963; Filler et al. 1976; Gross and Neuhauser 1948; Mustard et al. 1969). Therapeutic consequences among our own patients are discussed here and compared with reports in the literature.

Patients

Among 59 children operated on for esophageal atresia in Vienna during the past 20 years there were five cases of tracheal stenosis of varying origin (see Table 1). Another nine cases were treated in Munich from 1982 to 1984 (Table 2).

Of the five cases presented in Table 1, case no. 3 had probable tracheal compression by the innominate artery, and in cases no. 4 and 5 the diagnosis was certain. Case no. 5 is of special interest, since an erosive ulcer of the trachea was observed after asphyctic spells had temporarily ceased following tracheostomy and intubation. The tracheal ulcer disappeared following antefixation and truncopexy, and tracheal findings normalized. In three of the nine Munich cases truncopexy was performed; the other children improved without surgery.

Case no. 4 from Table 1 may be considered in detail. V.R., born June 18, 1983, birth weight was 2700 g, operated on for esophageal atresia type IIIb according to Vogt. Closure of the esophagotracheal fistula and primary anastomosis were performed. He received artifical ventilation postoperatively due to respiratory insufficiency and right-sided tension pneumothorax (Fig. 1), which relapsed twice despite thoracic drainage. A tracheostomy was performed on the 11th post-

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Table 1. Tracheal stenoses following esophageal repair (patients of the Dept. of Pediatric Surgery of the Mautner-Markhof Kinderspital, Vienna)

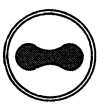
Patient, identification	Type acc. to Vogt	Therapy	Course	Findings	Form	Follow-up
1 PE, male, 1821	IIIb	Primary anastomosis, bougienage	Increasing cough after 4 weeks	Tracheal stenosis; fistula relapse after 5 weeks		Recurrent bronchitis
2 WM, male, 3802	IIIb	Primary anastomosis	Coughs after 1 year	Additional upper fistula; stenosis due to diverticulum		Normal tracheography and -scopy after 4 years
3 HR, male, 2799	IIIb	Gastrostomy on the 3rd day and ligation of fistula, esophagostomy without primary anastomosis impossible (4 weeks)	Cyanotic attacks after 2 months continuing despite tracheostomy	Tracheal stenosis above fistula; compression by innominate artery?		Death after 7 months; marasmus; bowel obstruction; necrotizing enterocolitis? Small-bowel fistulas!
4 VR, male, 17517	IIIb	Primary anastomosis	Asphyctic attacks increasing during feeding	Tracheal stenosis above fistula; compression by innominate artery; antefixation		Normal findings after 6 months
5 HF, male, 2412/76	IIIb	Primary anastomosis	Asphyctic attacks; tracheostomy; erosive ulcer	Tracheal stenosis above fistula; compression by innominate artery; antefixation		Normal findings after 1 month; death from bowel obstruction and sepsis later on

Table 2. Tracheal stenoses following esophageal repair (patients of the Pediatric Surgical Clinic, Dr. von Haunersches Kinderspital of the University of Munich, 1982–1984)

Patient, identification	Type acc. to Vogt; other diagnoses	Postoperative respiratory disturbance	Tracheoscopy; degree of stenosis	Combined angiotracheography	Therapy and course
1 RM, male, 6/30/78	IIIb; renal aplasia; deafness	Recurrent; therapy-resistant "bronchitis"	Pulsating TM, 90%	TC by innominate artery	TP suggested since symptoms unchanged
2 JS, female, 3/25/81	IIIb	Respiratory insufficiency; ventilator dependent	Pulsating TM, 90%	TC by innominate artery	Long-term ventilation, slow improvement after TP; stenosis of the right main bronchus later on
3 GA, male, 9/16/81	IIIb; lung aplasia; dextrocardia	Apneic spells; ventilator dependent	Pulsating TM, 90%	TC by innominate artery	Only closure of fistula, primary anastomosis impossible; clear improvement after TP
4 LR, male, 2/15/82	IIIb	Recurrent; therapy-resistant "bronchitis"	Pulsating TM, 80%	—	No essential complaints
5 HM, male, 2/20/83	IIIb	Apneic spells; reanimation; chronic "bronchitis"	TM, pulsating TC; 90%	TC by innominate artery	Slow improvement after TP
6 GD, female, 7/1/83	IIIb	Ventilator and CPAP dependent	TM, pulsating TC; 90%	TC; innominate artery? compression of left main bronchus	After 1 year additional intubation stenosis; very slow improvement

7	JS, male, 12/8/83	IIIb	Recurrent, therapy-resistant "bronchitis"	Pulsating TM, 90%	—	Improvement
8	KG, male, 14/8/83	IIIb, duodenal stenosis	Apneic spells; ventilator dependent	Pulsating TM, 70%	—	Slow improvement over months
9	KS, female, 31/10/83	IIIb; ASD, VSD, PDA	Apneic and cyanotic spells	TM, pulsating TC; 80%	TC in congenital heart disease	Fistula relapse requiring operative closure of fistula (following attempted endoscopic glueing); slow improvement after cardiac surgery (total correction)

Abbreviations: TM, tracheal malacia; TC, tracheal compression; TP, truncopexy; PDA, persistent ductus arteriosus

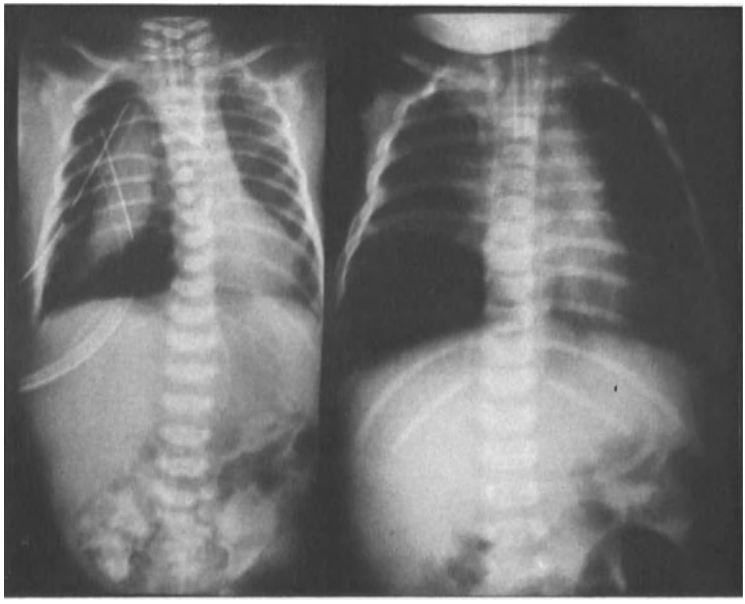


Fig. 1. Case no. 4, Table 1: *left,* spontaneous pneumothorax on 4th postoperative day, drainage; *right,* right lung emphysematous, 4 months postoperatively

operative day since the child could not be taken off the ventilator. Thereafter, oral feeding could be increased up to 8×50 g per day. Following attempted closure of the tracheostomy he had two cyanotic spells during feeding. Endoscopy revealed a transverse tracheal obstruction distal to the tracheal cannula (Fig. 2). With the tracheal cannula in place oral feeding could be continued, and the child was transferred to another hospital.

Because the infant was in good condition and did not exhibit any signs of respiratory insufficiency, the tracheal cannula was withdrawn. From this time on the patient developed recurrent severe asphyctic attacks which always appeared during feeding, thus necessitating further feeding via a gastric tube (Fig. 3). Then no asphyctic attacks occurred and the child thrived, but cyanotic spells always reappeared when oral feeding was tried. The patient was readmitted to our hospital on October 14, 1983, and an endoscopy performed on October 27, 1983 revealed a pulsating stenosis. Compression of the site of the stenosis effected pulse loss in the right forearm, whereas the pulse in the left forearm remained unchanged (Fig. 2). Thus, the stenosis was assumed to be due to the innominate artery. A second tracheostomy was performed on November 19, 1983 following reviewed attempts at oral feeding accompanied by severe cyanotic attacks. Thereafter oral feeding was possible. A new, severe attack required artificial ventilation. Angiography was therefore not possible. Computerized tomography disclosed a vessel immediately in front of the tracheal stenosis (Fig. 4). Surgical antefixation of the aorta and the innominate artery was performed on November 21, 1983; the tracheal cannula could be withdrawn on the 4th postoperative day.

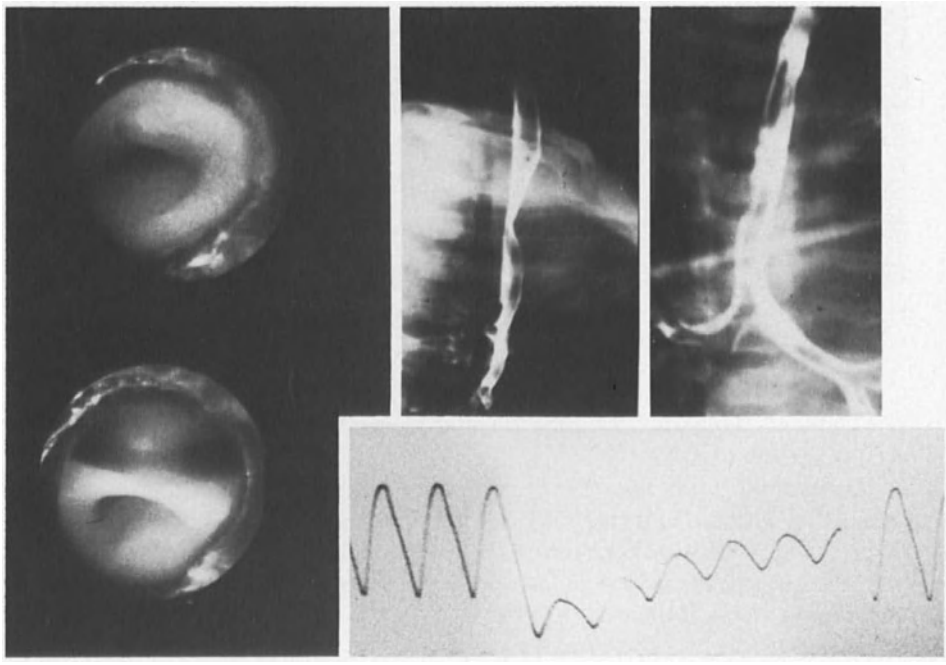


Fig. 2. Case no. 4, Table 1: *upper left,* endoscopic view of tracheal stenosis caused by innominate artery; *lower left,* distal to preceding view, trachea of normal width and typical dorsal esophagotracheal fistula; *upper right,* anterior tracheal compression caused by innominate artery; *lower right,* pulse tracing of right forearm during intratracheal compression of stenotic region by endoscope

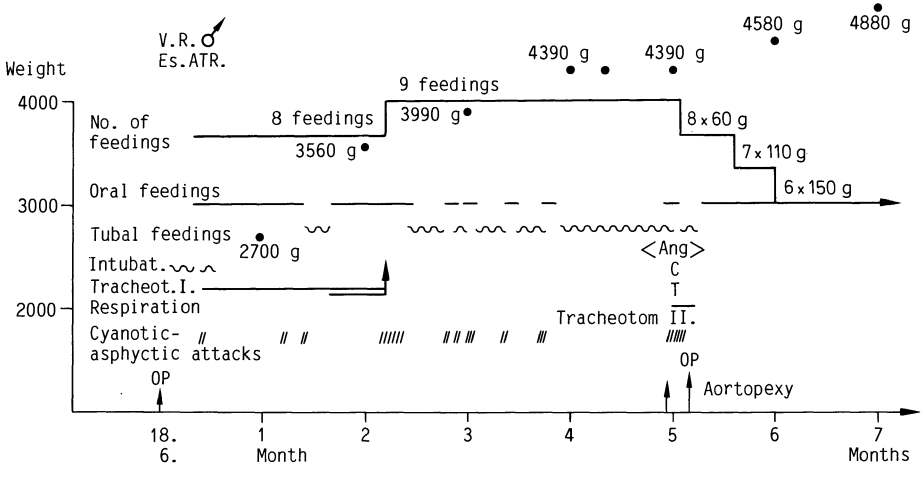


Fig. 3. Case no. 4, Table 1: course of feeding program and asphyctic attacks. —— Oral feeding; ~~~~ tubal feeding; /// cyanotic-asphyctic attacks

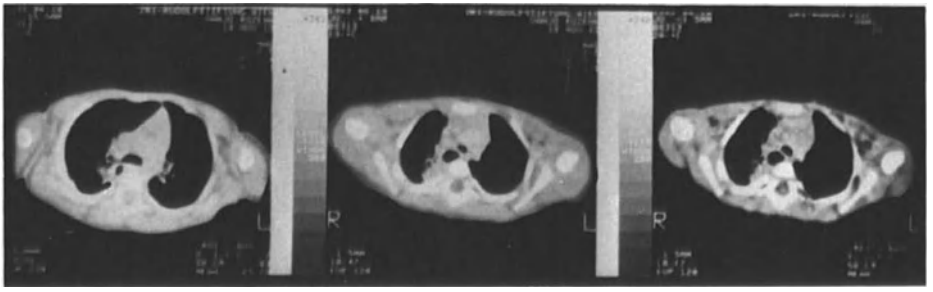


Fig. 4. Case no. 4, Table 1: CT scan with contrast medium for vessel exhibition. ⇳ Tracheal stenosis; ↗ innominate artery

The child was discharged from the hospital 20 days following surgery, with 6 × 150 ml meals given orally.

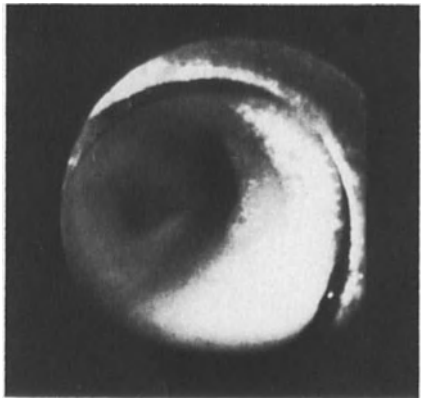
There had been at least 15 severe cyanotic and apneic spells, partly of a life-threatening character, finally requiring continuous artificial ventilation. Interestingly, the cyanotic attacks occurred only when the child was not tracheostomized and was fed orally. Control endoscopy 2 months postoperatively did not disclose any signs of tracheal stenosis (Fig. 5).

Discussion

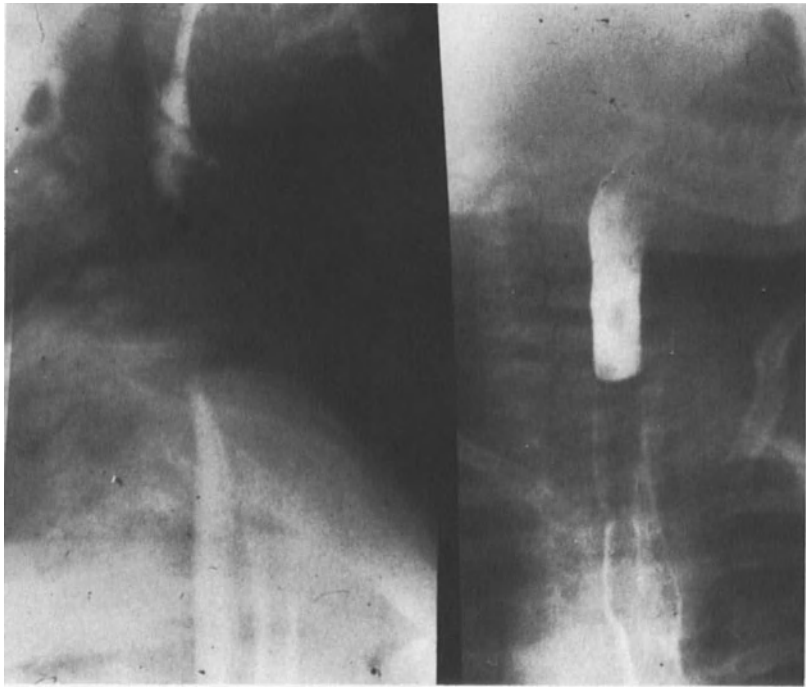
Based on our experience, we conclude that tracheal stenosis might well be responsible for respiratory disturbances following operations for esophageal atresia. Thus, postoperative endoscopic investigations are necessary. Failure to thrive following operations for esophageal atresia might also be due to tracheal stenosis.

Cases no. 4 and 5 of Table 1 and those in Table 2 show that the characteristic symptoms of respiratory disturbances may occur relatively late; symptoms seem to reach a maximum in the 3rd month of life (Filler et al. 1976; Mantel et al. 1979; our own cases). In severe cases (eight of our own cases) the complaints increase sharply, rendering urgent surgery necessary. Above all, fistula relapse must be excluded endoscopically in these cases. Additional upper esophagotracheal fistulas and large pseudodiverticula of former fistulas must be disproved, too (Daum 1970). Moreover, gastroesophageal reflux and disturbed esophageal motoric function are also to be considered. Endoscopic diagnosis of a pulsating, transverse tracheal obstruction with tracheal malacia makes compression by the innominate artery highly probable. This can be proven if decrease or loss of pulse occurs in the right forearm when pressure to the stenotic region is exerted against the thoracic wall (Fig. 2).

Diagnosis is made by means of simultaneous angiography and tracheography (Mantel et al. 1979; Smalhout and Hill-Vaughan 1979), which may be risky, however, in a child with respiratory insufficiency. In case no. 4 angiography could not be performed due to severe cyanotic attacks. An alternative is computerized tomography with contrast medium for vessel exhibition (Fig. 4).



a



b c

Fig. 5a–c. Case no. 4, Table 1: **a** normal findings during endoscopy 2 months postoperatively; **b, c** tracheography shows tracheal stenosis has disappeared

Operative truncopexy or antefixation of the innominate artery improves the situation; however, symptoms may not disappear at once, but only gradually, after a certain latency period. Formerly, vagovagal or central nervous reflexes were assumed to be the cause of cyanotic or bronchitic attacks after surgery for esophageal atresia; this has now been abandoned (Filler et al. 1976). The assumption of idiopathic tracheal malacia is also not valid in our opinion (Benjamin et al.

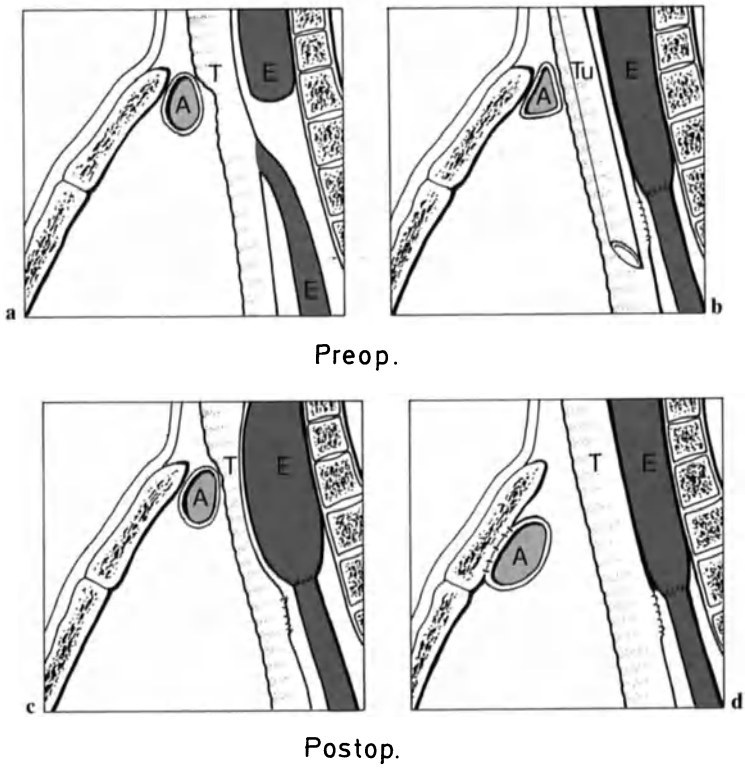


Fig. 6a–d. Position of esophagus *(E)*, trachea *(T)*, and innominate artery *(A)* in esophageal atresia: **a** before and **b** after esophageal anastomosis; **c** before aortopexy; **d** after aortopexy

1976; Emery and Haddadin 1971). But we agree that tracheal growth is impeded by the large upper esophageal pouch (Davies and Cywes 1978) and that the trachea is compressed between the upper pouch and the innominate artery (Filler et al. 1976). Following surgery the compression may be exacerbated, since the upper pouch, which was more cranial before, is placed into the narrow thoracic aperture (Fig. 6a, preoperative; Fig. 6b, c, postoperative).

The structure best suited to give way is the trachea. Particularly during swallowing, the trachea is displaced against the innominate artery, leading to respiratory obstruction (Fig. 6c). Antefixation with slight upward traction of the innominate artery provides more space for the trachea, thus avoiding respiratory obstruction. This mechanism was proven in eight cases of Filler's series (Filler et al. 1976) and two of our own. Contrary to other compressions of vascular origin with permanent stridor, cyanotic attacks occur predominantly during feeding in these children, who are otherwise relatively symptom free.

Tracheostomy with cannulation of the stenosis leads to temporary cessation of symptoms (two of our cases; Fig. 6b), but it should not be used as a permanent measure since tracheal malacia and erosive bleeding may result owing to bruishing

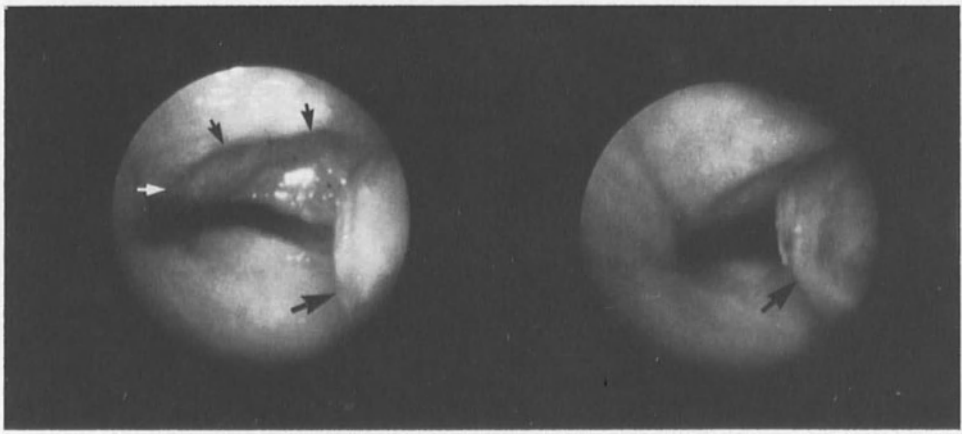


Fig. 7. Case 5, Table 1: *left,* tracheal stenosis above fistula following attempted pertubation with long cannula; erosive ulcer ↗; later antefixation of innominate artery. Withdrawn ⇳ cannula visible at the right side

of the tracheal wall between the cannula and the innominate artery (case no. 5, Table 1; Fig. 7). The tracheal cannula should therefore be withdrawn as early as possible.

The incidence of tracheal stenosis in esophageal atresia is reported to be 1 in 40 by Filler (Filler et al. 1976), which is roughly in accordance with our own figures. It is a very dangerous condition, leading to asphyctic attacks during feeding.

Summary

The significance of tracheal stenosis in children operated upon for esophageal atresia is described. Stenosis is caused mainly by tracheal compression by the innominate artery leading to tracheal malacia. Following esophageal repair tracheal compression is increased causing life-threatening asphyctic attacks during feeding. After establishment of diagnosis urgent surgery by means of truncopexy is indicated. The incidence of this complication is about 1 in 20 according to our figures and 1 in 40 according to Filler (Filler et al. 1976).

Résumé

Les auteurs étudient la signification d'une sténose trachéale chez les enfants opérés pour atrésie œsophagienne. La sténose est due à une compression par le tronc artériel innominé conduisant à une trachéomalacie. Après réparation de l'œsophage, la compression trachéale est passablement augmentée, entraînant des attaques asphyxiques compromettant l'existence pendant l'alimentation. Une fois le diagnostic établi, un traitement chirurgical urgent est nécessaire par réalisa-

tion d'une troncopexie du tronc innominé. L'incident de cette complication est d'environ de 1 pour 20 d'après nos constatations et de 1 pour 40 d'après celles de Filler.

Zusammenfassung

Es wird über die Bedeutung der Trachealstenose bei operierten Ösophagusatresien berichtet, wobei die wesentlichste Ursache eine Kompression der Trachea durch die A. anonyma sein dürfte, die eine Tracheomalazie bedingt. Durch die anatomischen Verhältnisse wird nach der Rekonstruktion des Ösophagus die Kompression der Trachea verschärft, die insbesondere während der oralen Ernährung bedrohliche Anoxieanfälle erzeugt. Die dringliche Therapie durch Trunkopexie ist daher nach Diagnosesicherung angezeigt; die Häufigkeit dieser Komplikation dürfte nach unserem Krankengut 3 auf 60 Fälle ausmachen, nach Filler ungefähr 1 auf 40.

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Colonic Interposition

N. V. FREEMAN[1]

History

In 1894, Bircher was the first to use a multistage antethoracic skin tube to replace the esophagus. The first babies to survive with esophageal atresia underwent similar multistage procedures following gastrostomy and cervical esophagostomy (Leven 1936; Ladd 1944). Jejunum was used via the antethoracic subcutaneous route by Roux (1907) and by Swenson (1947) for esophageal atresia. In one case of esophageal atresia the stomach was used via an anterior subcutaneous route, and later placed intrathoracically by cleavage of the underlying tissues (Potts 1950). A gastric tube was advocated by Jianu (1912) and used in esophageal atresia by Boerema (1951). The colon was used by Kelling (1911) and then by Lundblad for a 3-year-old with a caustic stricture (1921). Colon interposition via an intrathoracic route was first attempted by Sandblom in 1948 (Sandblom 1948), and Waterston (1972) was the first to use a graft of transverse colon based on the left colic artery, via the diaphragm and the left chest, through the apex of the pleura to join the cervical esophagotomy. The retrosternal route with the right colon was used by Javid (1954).

In 1970 a modification of the Waterston technique was used by the author (Freeman and Cass 1982). The main difference is that the colon is placed in the normal esophageal site in the posterior mediastinum, mainly extrapleurally, and not intrapleurally, in the left chest behind the left lung (Fig. 1). The Waterston operation was further modified by passing the colon via the esophageal hiatus after removing the lower esophageal stump, and anastomosing the lower stump to the posterior wall of the stomach.

Patients and Method

During the past 18 years I have performed 34 colon transplants; 19 were via the normal esophageal route, five were retrosternal, and two were Waterston operations. Seven were performed in the neonatal period or early infancy. During this period I have operated on just over 100 cases of esophageal atresia. A primary anastomosis was possible in 77% of the cases, but 26 required colonic interposition. Nine of the 26 cases of esophageal atresia were type IIIb (Vogt) performed under tension with subsequent disruption. Ten were type IIIa with a wide gap.

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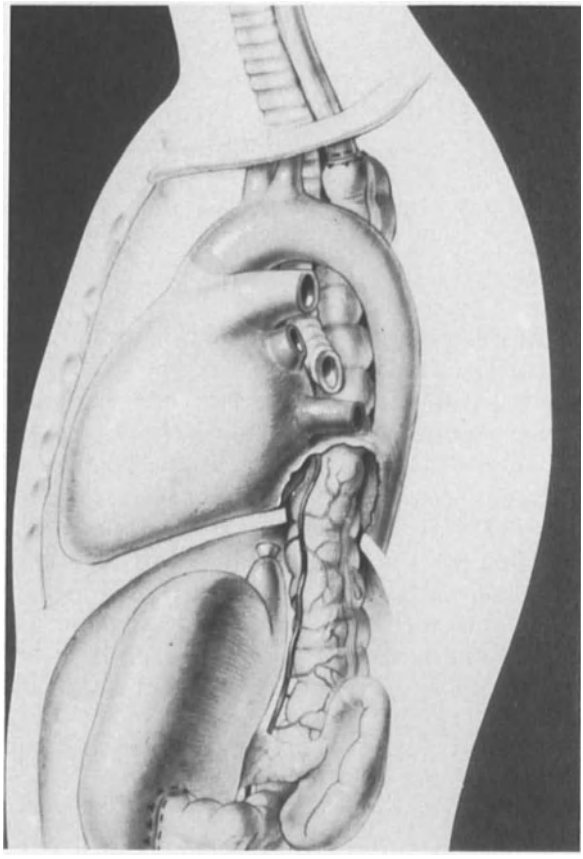

Fig. 1. Modification of Waterston technique used by Freeman (Freeman and Cass 1982); colon placed in normal esophageal site

Seven cases were pure esophageal atresia without a fistula. The rest of the interpositions were for caustic or peptic strictures, referred from elsewhere or operated on in the patients' own hospitals for teaching purposes.

Evolution of the Current Method of Treatment

In 1961, while working with Waterston at Great Ormond Street, I was impressed with the difficulties experienced during the 'blind' dissection of the thoracic inlet and dilatation of the tunnel for the siting of the graft. Strictures occurred at the upper anastomosis, and it was not usually possible to pass a rigid esophagoscope through it. Dilatations had to be performed 'blind', using gum elastic bougies. Today, using a flexible endoscope, dilatations are easier.

Route

Waterston considered it important to leave the lower esophageal stump in situ to prevent gastrocolic reflux (Azar et al. 1971), and to perform the lower anastomosis

in the left chest using a paraesophageal tunnel for the colon and vascular predicle. The vagus nerves are preserved by this manoeuver. He believes that if the distal esophagus is preserved the cardioesophageal sphincter remains intact, reflux of acid is prevented, and the act of deglutition is followed by relaxation of the cardio-esophageal junction. He states, however, that in case of peptic stricture, when the lower esophagus is replaced by a long intra-abdominal length of colon into the posterior wall of the stomach, reflux of gastric juice and air into the colon are prevented, and that peptic ulceration and tortuosity of the graft does not take place. This has not been my experience.

As the graft is based on the upper left colic artery, it is difficult to shorten the descending colon enough to prevent redundancy in the left chest. The colon graft can be shortened by careful division of the arcades and removal of a portion of the distal colon, but this was not tried. The natural lie of the graft based on the upper left colic artery is through the hiatus, placing the pedicle, but not the graft, behind the pancreas and entering the posterior wall of the stomach. This method was used in about half the cases.

The retrosternal route was used in five occasions, two were for caustic strictures. Again the major disadvantage was kinking in the neck, and there were postoperative difficulties in trying to dilate or to see the upper esophagus-colon anastomosis.

Optimal Age for Operation

Waterston (1972) favuored 6–12 months as the optimal age for colonic interposition; in all his cases the operation was preceded by a cervical esophagostomy. There are two disadvantages to colon interposition at this age: (a) the baby is not really taking solid food, which would help to dilate the anastomosis, and (b) the

Table 1. Complications arising in the 19 cases in which the normal esophageal bed was used

Laryngeal nerve damage	3
Cervical leak	2
Upper-end stricture	2
Lower-end stricture	2
Redundant colon (later resection)	2
Stenosis of cervical esophagostomy	1
Gastrostomy leak	6
Intestinal obstruction (pyloric)	1
Hematemesis	1
Severe wound infection (osteomyelitis)	1
Failure to thrive	2
Technical failure to develop tunnel	1
Tracheostomy	1

Table 2. Data on six patients who underwent early colonic interposition

Case	Date of birth	Gestation (weeks)	Birth weight (kg)	Age at operation (weeks)	Weight at operation (kg)	Complications	Current weight (percentile)	Postop. time to discharge
1. NMcG	10/06/74	40	2.85	4	3.3	Loss of graft	† 7 days postop.	†
2. ST	03/09/79	36	2.32	8	3.7	Upper leak FTT[a]; now well	50	2½ months
3. HE	22/11/79	34	1.6	16	4.2	None	25	22 days
4. MB	14/01/81	32	1.45	20	3.8	Marked FTT[a]; late stricture (1 year postop.) at gastrocolic anastomosis	$\ll 3$	10 months
5. AC	05/05/81	40	3.64	2	3.7	None	20	21 days
6. GC	26/03/83	36	2.50	2	2.6	None	<3	9 days

[a] FTT = Failure to thrive

thoracic inlet is small, and finger dissection, or pulling up a bulky colon through this area, may lead to postoperative respiratory difficulties and possible laryngeal nerve damage. For these reasons I preferred to wait until the child is about 18 months old for the colonic interposition, as by then the baby should be well established on solid food.

This policy has recently been modified. In the past, neonatal colonic interposition was considered too risky (Petterson 1962; Bentley 1965; Soave 1972). Because of the possible morbidity with cervical esophagostomy (see Table 1) it was not performed in the last five cases. Replogle tube drainage of the upper pouch was carried out from 2 weeks to 4 months, and early interposition performed. The technique in the neonatal period consists of an immediate gastrostomy and Replogle tube drainage. At about 2 weeks of age or sooner the neonatal colonic interposition is carried out. The patient is positioned lying on his or her left side so that the epigastrium and right axilla are exposed. A transverse-muscle cutting incision with a vertical midline extension and freeing of the gastrostomy is carried out. A short graft of transverse colon based on the upper left colic artery is fashioned. A Denis Browne extrapleural approach to the upper esophageal segment is made. Extrapleural finger dissection to the hiatus, from above and from the abdomen, develops a tunnel through which the graft is pulled up and anastomosed to the upper esophagus. The distal colon is anastomosed to the posterior wall of the stomach. The results in this group are very encouraging, and at present this is my method of choice in management of long-gap esophageal atresia (Table 2). Figure 2 shows the ages at operation in the author's personal series. Note the trend towards a neonatal operation in the past 8 years.

Results

The success of surgery may be difficult to judge, but several parameters are useful.

Morbidity

The main complications in the series of neonatal posterior mediastinal colon transplants are shown in Table 1. The earlier patients were treated in Liverpool, and a full follow-up of these cases has not been possible.

Associated Anomalies

Anomalies are not common in this group, but when present they contribute to the mortality and mordidity (Table 3). One case of mental subnormality was acquired at 4 months when the baby suffered an anoxic episode during prolonged fitting, which was not treated promptly.

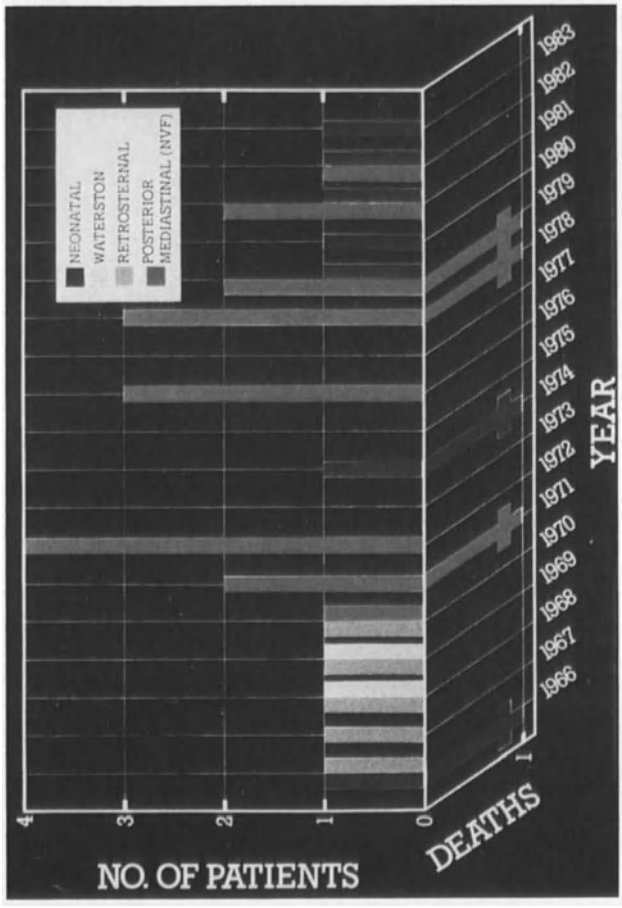

Fig. 2. Techniques employed in author's series at operation

Table 3. Anomalies in patients associated with esophageal atresia

Hydrocephalus	1
Optic atrophy	1
Single kidney + megaureter	1
Annular pancreas	1
Mental subnormality	4

Mortality

The patient with hydrocephalus died 2 days after revision of her value for raised intracranial pressure. The first neonatal colonic interposition was performed in 1966 and was a modification, in that the transverse colon was joined to the upper

esophagus but the lower colon was left in situ. The child regurgitated feces and died. There have been no neonatal deaths among the last five cases. The death in 1979 shown in Fig. 2 occurred 3 years after the colonic interposition, from tracheal stenosis following tracheostomy.

Dysphagia

An assessment of the eating habits of the child is difficult. One way is to compare him with a younger or older sibling regarding the type of food he eats and the length of time he takes to complete a meal. In the "good" cases there is no appreciable difference. In those failing to thrive, it appears that the child is unable to eat a large meal, and frequent small meals are more acceptable. Recently, Andrassey et al. (1983) assessed the nutritional status of 51 esophageal atresia patients. No significant difference was found between those with a birth weight of

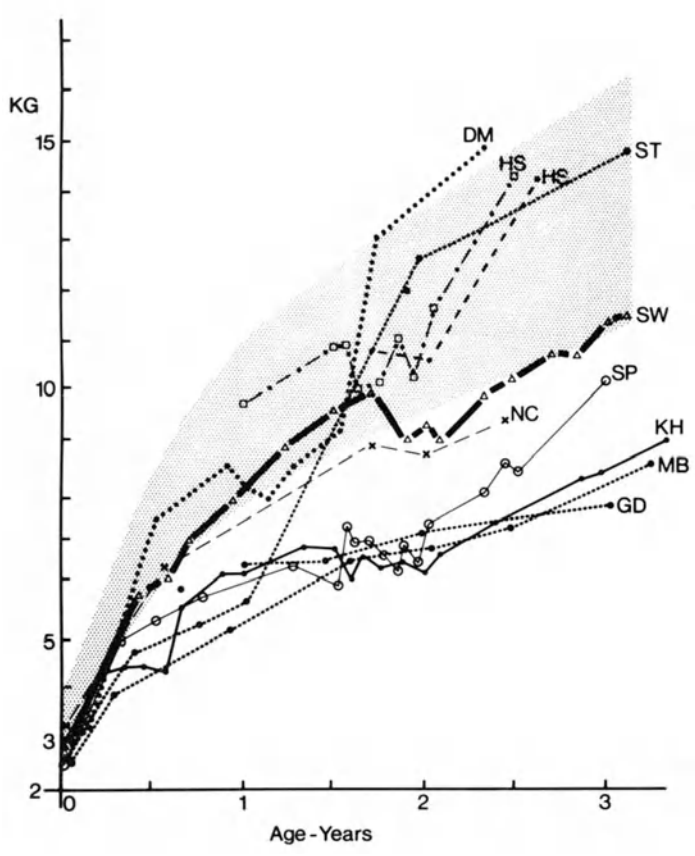

Fig. 3. Weights of children following colon interposition. Note the large majority who run along the 3rd percentile for weight, and note in particular the graphs of S.T., M.B., and G.D. (see text)

less than 2500 g and those over 2500 g; 19% remained below the third percentile for height and weight.

Three cases require special mention (Fig. 3): G.D. appeared to be suffering from calorie deprivation. On a careful, balanced study he was found to be taking in 1160 kcal/day instead of the recommended 1560 kcal for a 2- to 4-year-old child.

M.B. appeared to be failing to thrive but he is genetically small, as his parents are 140 cm and 155 cm tall. He is in the 50th percentile for his expected height and weight.

There is clear evidence that psychosocial stress causes relative failure to grow by inhibiting the secretion of growth hormone (Tanner 1978). The growth curve of S.T. demonstrates a dramatic increase in weight once he was taken into foster care.

Redundancy and Kinking

The most serious problem remains redundancy and kinking. The etiology is not clear, but obstruction at the hiatus or lower anastomosis is probably significant, as redundancy is not commonly seen in adults. A modification as described by Lynn (1973) was used in an attempt to prevent this redundancy. In the last case a very short graft was taken, and the longitudinal teniae were divided to the mucosa to straighten and lengthen the graft (Fig. 4). The postoperative barium study showed

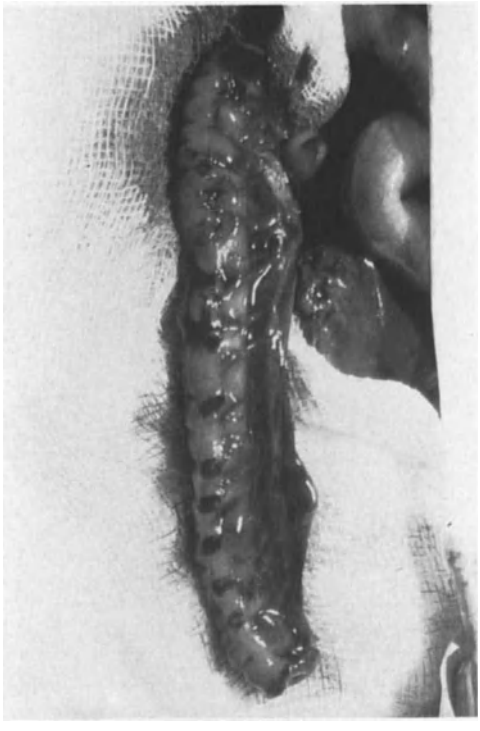

Fig. 4. Short colon graft with tenia divided to straighten the graft

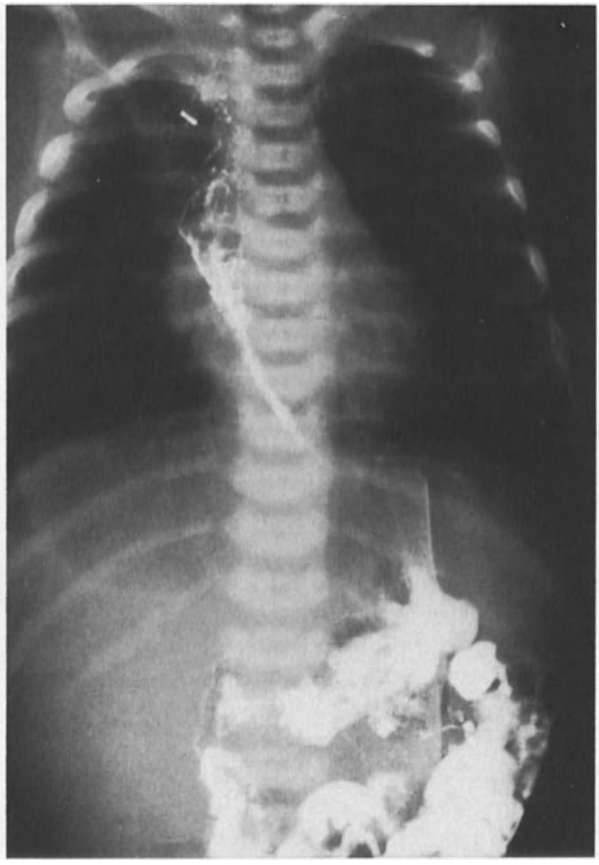

Fig. 5. Barium study at 3 months showing graft to be straight, with no redundancy, following division of tenia

the colon to be straight, with no kinking (Fig. 5). This modification has so far, with a short follow-up of 9 months, been successful.

Discussion

Functional studies using barium, cineradiology, and manometry show that the colon graft acts as a passive conduit and that reflux is common.

Sieber and Sieber (1968) studied ten patients with either a retrosternal or Waterston-type interposition. During cineradiography the width increased and the haustra became more distinct. Distal emptying was slow with the patient recumbent and rapid when he was erect. Transit time from deglutition to entrance into stomach varied from 18 s to 5 min. Prolonged observations in two cases revealed ineffectual contractions with swallowing. No reflux was seen in the four Waterston cases. Marked redundancy was noted in two cases. Intraluminar manometric pressure studies using a triple lumen tube revealed a normal upper esophagus, but in all patients studied the colon was essentially without muscular

activity. The only waves noted were related to a bolus pushed by the active esophagus from above.

Schiller, Frye, and Boles (1971) studied 29 patients. Barium studies showed prompt passage into the graft, and no peristalsis was seen in any study; the segments fill and empty by gravity. Passage into the stomach was prompt in 18 cases and slow in six. Redundancy was mild in 15 and marked in nine. Some correlation was noted between redundancy and delayed emptying. No differences in emptying or clinical evaluation were noted between iso- or antiperistaltic grafts. The authors conclude that the grafts function as passive conduits.

Rogers et al. (1978) studied 13 children. Most had a right colonic interposition with the terminal ileum attached. In three with more than 10 cm of ileum the Vit B_{12} status was abnormal. The ileum maintained normal peristalsis but the colon showed no coordinated, and only occasional random peristalsis. With three patients a thick paste reached the stomach in less than 3 min; it took more than 30 min with another three patients. When liquid was offered there was prompt entry into the stomach.

Belsey (1983) states:

> Vulliet promulgated a heresy that since colon does not normally exhibit peristalsis, anti-peristaltic transplants should prove as effective as isoperistaltic transplants. This fallacious reasoning, ignoring the unidirectional propulsive activity of normal colon, peristaltic or otherwise, perpetuated the use of transverse colon and resulted in untold disability and disaster from complications of anti-peristaltic transplants.

A prospective study was carried out by the author (unpublished) of ten children with colonic interpositions using Tc99-labeled milk to determine the rate of emptying of the graft. The effect of gravity was studied with five patients who were tested both sitting and in a supine position. The results are shown in Table 4. In the supine position the food required to 14–30 min reach the stomach or did not do so until the patient was placed in the erect position and given a drink. Spontaneous reflux occurred on four occasions while the patient was sitting but in seven patients when they were positioned head down.

Conclusion

The successful bridging of the gap, whether large or small, between the upper and lower segments remains a formidable problem. The use of a colon graft remains a satisfactory method, in spite of a significant morbidity. There appears to be a distinct advantage with neonatal colonic interposition considering the low morbidity and mortality and a short hospital stay of 3–4 weeks in total.

Summary

Intrathoracic colonic interposition in children with esophageal atresia has been used for the past 36 years. The preferred routes have been via the left pleural cavity and retrosternally.

Table 4. Results of Tc99-labeled milk study comparing late with early (neonatal) colon interposition

Age at study	Sex	Erect	Supine	Emptying time	Spontaneous reflux	Positioned reflux	Stomach- and small-bowel activity
Late							
LR							
7 years	F	+		22 min	−	+	Free
8 years			+	Pooling in conduit, clearing with erect drink @ 25 min	−	+	Nil till drink 25 min onwards
NB							
6 years	M	+		<25 min	−	−	Free
6 years			+	Pooling in conduit	−	−	14 min onwards
FD							
11 years	M	+		20 min	−	−	Free
SW							
5 years	F	+		>45 min	−	+	12 min onwards
GD							
2 yr, 1 mo	M	+		>50 min	+	−	10 min onwards
2 yr, 10 mo			+	Pooling in conduit >3.5 h	−	−	30 min onwards
FB							
7 years	F	+		>22 min	+	−	Free

Table 4 (continued)

Age at study	Sex	Erect	Supine	Emptying time	Spontaneous reflux	Positioned reflux	Stomach- and small-bowel activity
Early							
HE							
3 years	F	+		15 min		+	Free
AC							
11 months	M	+		>1 h	+	+	Free
MB							
18 months	M	+		>45 min	+	+	15 min onwards
2½ years			+	Pooling in conduit, clearing with erect drink @ 25 min	—	—	Nil till drink, 25 min onwards
ST							
2 yr, 3 mo	M	+		<50 min	–	–	Free
3 yr, 5 mo			+	Pooling in conduit, clearing with erect drink	–	+	25 min onwards

Over the past 13 years the author has developed a method using the posterior mediastinum (normal esophageal route) for the colonic interposition. An analysis of 19 personal cases using this method is presented. A further nine cases using either the retrosternal or the Waterston routes and six cases using a modified neonatal operation are included.

The evolution of the method currently used by the author is traced. Major problems remain, and further modifications relating to the optimal timing for the colonic interposition, length of colon graft, and siting of the lower cologastric anastomosis continue to be tried and are discussed.

The function of the graft has been studied with ten patients using ^{99}Tc-labeled milk. The results of this study (unpublished) are presented. Gravity is shown to be the major factor influencing the onward passage of milk and food between the colon and stomach.

Résumé

Une œsophagoplastie colique intra-thoracique a été pratiquée chez l'enfant porteur d'atrésie œsophagienne pendant les dernières 36 années. Les abords utilisés ont été la cavité pleurale gauche et la voie rétrosternale.

Depuis 13 ans, les auteurs ont développé une technique utilisant le médiastin postérieur (voie normale de l'œsophage) pour l'interposition colique. Une analyse des 19 cas personnels utilisant cette technique est présentée. Une autre série de 9 cas utilisant soit la voie rétrosternale, soit la technique de Waterston, est présentée, de même qu'une série de 6 cas utilisant une opération néonatale modifiée.

L'évolution de la technique habituelle utilisée par l'auteur est retracée. Les problèmes majeurs qui restent à considérer sont en relation avec le moment optimum pour l'interposition colique, la longueur du greffon colique, l'emplacement de l'anastomose colo-gastrique basse. Tous ces éléments sont discutés.

La fonction de la greffe a été étudiée chez 10 patients en utilisant le ^{99}Tc mélangé à du lait. Les résultats de cette étude (non publiée) sont présentés. On démontre que la progression par gravité est le facteur majeur influençant la progression du lait et des aliments entre le côlon et l'estomac.

Zusammenfassung

In den letzten 36 Jahren wurde die intrathorakale Koloninterposition bei Kindern mit Ösophagusatresie durchgeführt, und zwar durch die linke Pleurahöhle oder retrosternal. In den letzten 13 Jahren wurde vom Autor für die Koloninterposition das hintere Mediastinum (normale Ösophaguslage) benutzt. Anhand von 19 persönlich operierten Fällen wird diese Methode dargestellt. Weitere 9 Fälle wurden entweder retrosternal oder nach der Methode von Waterston transpleural links operiert; 6mal wurde eine modifizierte Operation bei Neugeborenen ausgeführt. Die Entwicklung der derzeitigen Methode wird dargestellt; größere Pro-

bleme und weitere Modifikationen (bezogen auf den optimalen Zeitpunkt für die Koloninterposition, die Länge des verwendeten Koloninterponats und die Position der unteren kologastrischen Anastomose) müssen noch erprobt werden und werden diskutiert. 10mal wurde die Funktion des Interponats untersucht durch ^{99}Tc-markierte Milch. Die bisher nicht publizierten Ergebnisse dieser Studie werden mitgeteilt. Als Hauptfaktor für die Nahrungspassage zwischen Kolon und Magen ist die Schwerkraft anzusehen.

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Management of Long-gap Esophagus: Experience with End-to-End Anastomosis Under Maximal Tension

S. Hagberg[1], A. Rubenson[1], U. Sillén[1], and K. Werkmäster[1]

During the years 1969–1982, 98 patients with esophageal atresia underwent surgery at our institution. In 15 of the patients a distance of more than 2 cm between the esophageal ends was noted after maximal mobilization of the proximal and distal esophageal segments. Eight of these infants had long-gap atresia with tracheoesophageal fistuale (LGF) to the lower segment. In all these patients end-to-end anastomosis was performed in one layer after closing the tracheoesophageal fistula by the transpleural route, taking great care not to damage the circulation to the lower esophageal segment. A transanastomotic nasogastric tube was left in situ in all patients.

Anastomotic leakage was noted in all LGF patients, for a mean period of 20 days. It was treated conservatively in five patients with one or two pleural drainage catheters. Three infants had to be reoperated because of severe atelectatic changes in the right lung causing severe ventilatory problems. Postoperative esophagoscopy with calibration of the anastomosis was done in all patients. Anastomotic obstruction was found in six of the eight LGF patients and was treated with repeated esophageal dilatations. Gastroesophageal reflux was noted in three of the eight LGF patients postoperatively. One patient was treated conservatively and two had to undergo a fundoplication procedure. All eight LGF patients are now doing well with no or only minor swallowing problems.

The longest distances between the esophageal ends were found in patients with long-gap esophageal atresia but without fistulae (LGNF) (Fig. 1). The treatment difficulties with these patients are reflected by the various surgical procedures performed for esophagus correction. End-to-end anastomosis of the esophageal ends was performed in four cases, in two patients primarily and in two as a delayed operation after continuous salivary suction of the upper segment for 3 and 12 weeks respectively.

Of the two patients who were anastomosed primarily, one had to be reoperated because of severe anastomotic leakage but is now doing well after repeated esophageal dilatations. The other one also developed a leak and a severe anastomotic stricture. This latter patient died from complications due to esophageal dilatation (perforation, mediastinitis). One of the patients subjected to delayed anastomosis recovered uneventfully except for minor leakage and gastroesophageal reflux, which disappeared after conservative therapy. However, the other patient subjected to delayed esophageal anastomosis developed an intractable esophageal stricture which had to be reoperated twice, one and four years after the primary

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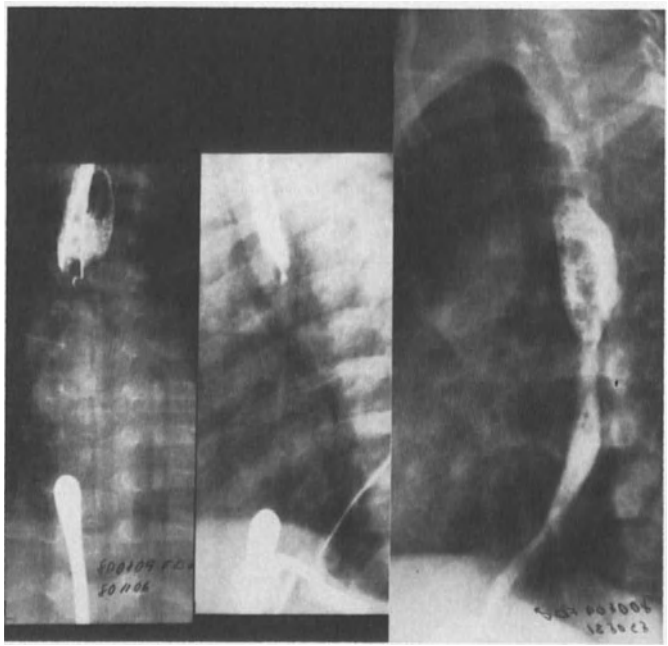

Fig. 1. Pre- *(left, middle)* and post- *(right)* operative X-ray films of an infant with long-gap esophageal atresia without fistula (a metal tube is inserted in the lower segment through the gastrostoma), operated upon with a delayed end-to-end anastomosis at the age of 12 weeks

anastomosis. He had also had severe problems with esophageal reflux and has undergone fundoplication twice. However, now at the age of 5 years he is doing well without major swallowing problems.

Colonic transposition was done in the three remaining cases, two primarily and one delayed, according to Waterston. One of the babies subjected to primary colonic transposition died in the early postoperative period from cerebral hemorrhage. The other developed a stricture in the transposed colon segment postoperatively; it was treated with repeated esophageal dilatations without success. At exploration at the age of 14 months, total fibrosis of the colon segment was noted. However, direct anastomosis of the esophageal ends could now be accomplished under tension. This anastomosis restrictured and had to be resected. The patient is now doing well, without any swallowing problems. The patient subjected to delayed colonic transposition also developed stricture in the anastomosis between the colon and the esophagus. The obstruction had to be treated with a special dilatation procedure (steel olives over a transanastomotic nylon thread). This girl, now 6 years of age, is satisfied with her esophageal substitute except for minor swallowing difficulties.

Table 1. Surgical procedures used for correction of long-gap esophageal atresia, management of postoperative complications, present status

Surgical procedure	Esophageal atresia with fistula ($n = 8$)	Esophageal atresia without fistula ($n = 7$) (2 †)			
	End-to-end anastomosis ($n = 8$)	Primary end-to-end anastomosis ($n = 2$)	Delayed end-to-end anastomosis ($n = 2$)	Primary colonic transposition ($n = 2$)	Delayed colonic transposition ($n = 1$)
Leakage	8	2	2		
Treatment					
Drain	5	1	2		
Reoperation	3	1			
Stenosis	6	1	1	1	
Treatment					
Dilatation	6	1	1	1	1
Resection			1	1	
Gastroesophageal reflux	3		2		
Treatment					
Conservative	1		1		
Fundoplication	2		1		
Present status					
No or minor swallowing problems	8	1	2	1	1
Major swallowing problems					

Conclusion

Long-gap esophageal atresia presents difficult therapeutic problems. Different methods of treatment have been suggested. The policy in our unit has been to try to correct the esophagus with a primary end-to-end anastomosis under great tension, although we are aware that this procedure will often lead to anastomotic complications such as leakage, stricture, and sometimes brachyesophagus with gastroesophageal reflux. End-to-end anastomosis under great tension appears to be possible in most patients with long-gap esophagus with fistulae from the lower segment. However, in long-gap esophagus without fistulae to the lower segment, the distance between the ends may be too large to permit direct primary anastomosis. In our opinion, delayed end-to-end anastomosis under great tension is possible in

most of these patients. The postoperative complications are similar to those seen after primary end-to-end anastomosis. When delayed anastomosis is impossible, an esophageal substitute operation has to be performed. It has been suggested that myotomy of the upper esophageal segment should make an anastomosis feasible in long-gap esophagus. However, in the authors' opinion, this procedure combined with an end-to-end anastomosis under maximal tension is associated with too great a risk of esophageal rupture and impaired healing at the anastomotic level, and it has therefore not been applied in our unit. However, we do believe that myotomy might be beneficial in anastomosis with moderate tension, to reduce healing complications.

Summary

From 1969 to 1982, 15 cases of long-gap esophageal atresia were treated. Twelve patients could be managed by direct end-to-end anastomosis – ten primary and two delayed. Anastomotic leakage was noted in all patients, causing anastomotic obstruction in nine, which had to be treated with repeated esophageal dilatations. In one patient the esophageal dilatation resulted in esophageal perforation, with a fatal outcome. The remaining 11 patients are all doing well.

Three patients were subjected to colonic transposition, two primary and one delayed according to Waterston. One of these subjected to primary colonic transposition died postoperatively from cerebral hemorrhage. The other one is now doing well after resection of the transposed colonic segment which fibrotized but could be resected, after which the esophageal ends could be anastomosed. The patient subjected to delayed colon transposition is also free of major swallowing problems following immediate postoperative anastomotic stricture which could be managed by repeated dilatations.

Résumé

Les auteurs ont observé 15 cas d'atrésie œsophagienne "long-gap" entre 1969 et 1982. Douze patients durent être traités par anastomose termino-terminale directe. Chez 10, il s'agissait d'une anastomose primaire et chez 2 d'une anastomose différée. Chez tous les patients, on observa la production d'une déhiscence de l'anastomose qui conduisit 9 fois à une sténose dont le traitement consista en dilatations répétées; 1 fois la dilatation de l'œsophage entraîna une perforation avec issue fatale. Les 11 patients restants présentèrent une satisfaisante.

Chez 3 patients, on fit une interposition colique, 2 fois primitive et 1 fois différée selon Waterston. Un patient, chez qui une interposition colique primaire avait été pratiquée, mourut dans les suites post-opératoires d'hémorragie cérébrale, l'autre patient est en bon état général après résection du segment colique fibrosé transposé. On put faire une anastomose directe termino-terminale de l'œsophage après résection de ce segment. Le malade avec transposition colique différée n'a

pas de problèmes graves de déglutition après qu'une sténose anastomotique post-opératoire ait été traitée par dilatations répétées.

Zusammenfassung

Von 1969–1982 wurden 15 Fälle von langstreckigen Ösophagusatresien beobachtet. 12 Patienten konnten durch direkte End-zu-End-Anastomose behandelt werden; 10mal wurde eine Primäranastomose und 2mal eine aufgeschobene Anastomose durchgeführt. Bei allen Patienten kam es zur Anastomosendehiszenz, die in 9 Fällen eine Stenose erzeugte, welche mit wiederholten Dilatationen behandelt werden mußte. Einmal kam es bei der Bougierung des Ösophagus zur Perforation des Ösophagus mit tödlichem Ausgang. In den übrigen 11 Fällen war das Ergebnis gut.

Bei 3 Patienten wurde eine Koloninterposition vorgenommen, 2mal primär und einmal als aufgeschobene Operation nach Waterston. Ein Patient, bei dem die primäre Koloninterposition durchgeführt wurde, starb postoperativ an einer Hirnblutung, der andere Patient ist – nach Resektion des fibrosierten transponierten Kolonsegments – in gutem Zustand. Nach Resektion dieses Segments ließ sich eine direkte End-zu-End-Anastomose des Ösophagus anlegen. Auch der Patient mit aufgeschobener Kolontransposition hat keine schwereren Schluckprobleme mehr, nachdem eine unmittelbar postoperativ aufgetretene Anastomosenstriktur durch wiederholte Dilatationen erfolgreich behandelt werden konnte.

Experiences in the Treatment of Esophageal Atresia with Rehbein's Olive Technique

H. SAUER[1] and R. KURZ[2]

Today, esophageal atresia with lower fistula does not present major problems and can be treated successfully, even in prematures of group C according to Waterston. Difficulties arise, however, from long-gap esophageal atresia where primary anastomosis is not possible. In the majority of these cases lower fistulas are absent. Improved intensive care makes the long-term care of such children possible, even without salivation drainage via cervical esophagostomy. Waterston's colonic interposition, formerly accepted with enthusiasm, is being increasingly driven out by elongation methods which eventually allow for primary anastomosis. In 1965 Howard and Myers introduced elongation bougienage, for which Hendren developed an electromagnetic technique in 1975 (Hendren and Hale 1975). Livaditis advocated esophagomyotomy for elongation of the upper pouch in 1972 (Livaditis et al. 1972).

As early as 1954, Rehbein tried to connect the pouches by means of a catheter (Charrière 8) in long-gap esophageal atresia (Rehbein 1976). Five children thus treated, however, had lower esophagotracheal fistula, and four then developed fistula relapse and died. This method was therefore abandoned. In 1971 Rehbein published his method of elongation bougienage by means of metal olives and a thread running through the mediastinum (Rehbein and Schweder 1971). Thoracotomy became unnecessary with the introduction of endoscopic thread insertion by Okmian in 1975 (Okmian et al. 1975).

Since 1979, four children with esophageal atresia without lower fistula have been treated by Rehbein's thread-and-olive method at the Pediatric Surgical Clinic of the University of Graz. Two of these cases were presented together with Booß at the BAPS Congress in Oxford (Booß et al. 1982). The number of cases itself is not very impressive. However, time has come to subject this method to a critical analysis, even if only a few cases have been treated.

Case Reports

Case 1

H.J., male, born Jan. 15, 1979, 2900 g, 49 cm. Diagnosis of esophageal atresia was made by probing of the esophagus and a plain chest X-ray. Tracheoscopy and

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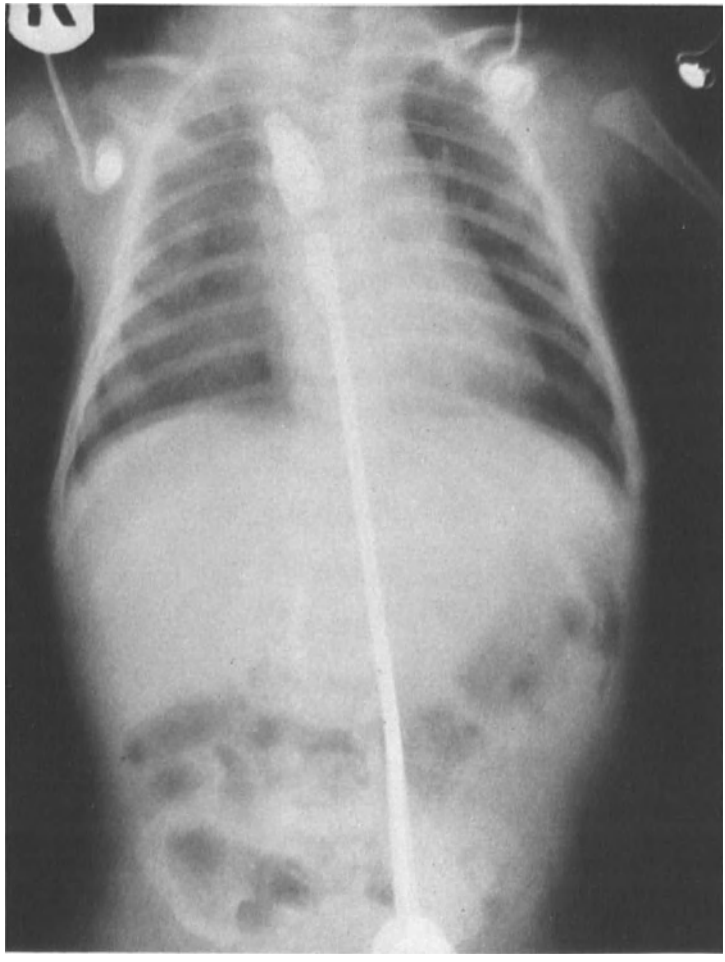
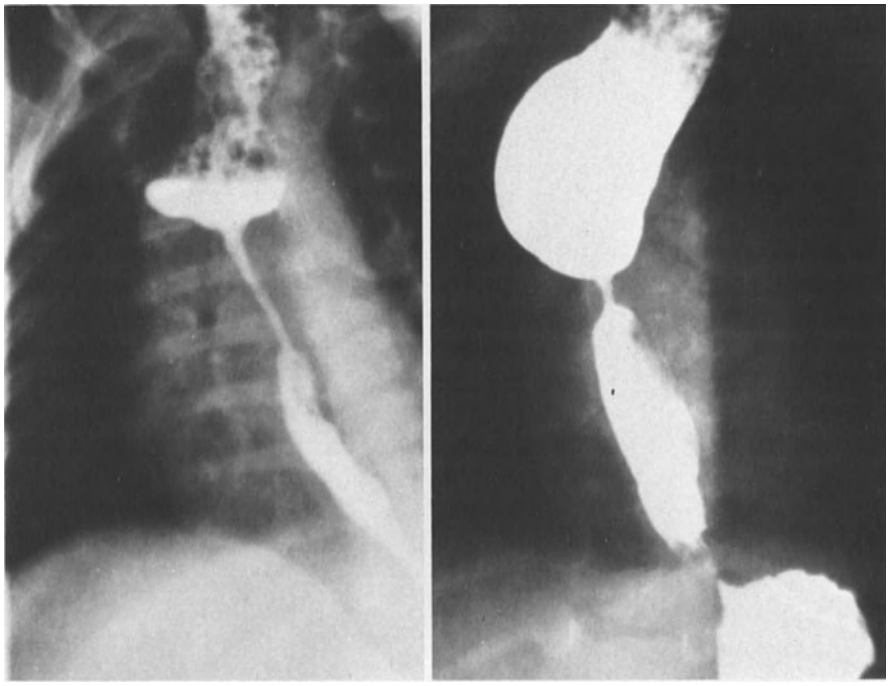
esophagoscopy (Jan. 16) revealed an upper esophagotracheal fistula. Since the pharynx and the esophagus were *Pseudomonas* contaminated and the child presented with septic signs, closure of fistula was postponed. Gastrostomy was performed on January 23. A bougienage thread was endoscopically inserted on March 3 by entering a Charrière-8 cystoscope into the lower pouch and an esophagoscope into the upper pouch. After radiographic control of the endoscope position, a monofile thread was advanced in a caudocranial direction. Finally, the esophagotracheal fistula was closed with double-row sutures and the esophageal site of the fistula covered with a pediculated muscle flap originating from the anterior cervical muscles. Bougienage with olives was started on the first postoperative day (Fig. 1a) and repeated seven times. From the 14th postoperative



a

Fig. 1a–f. H.J., born 15/1/79. **a** Double exposure: olives in resting position and under traction, 1 week following start of bougienage (March 3); **b** esophageal passage on March 23; **c** on May 30; **d** on August 10; **e** on December 19; **f** final findings on March 25, 1980



b c

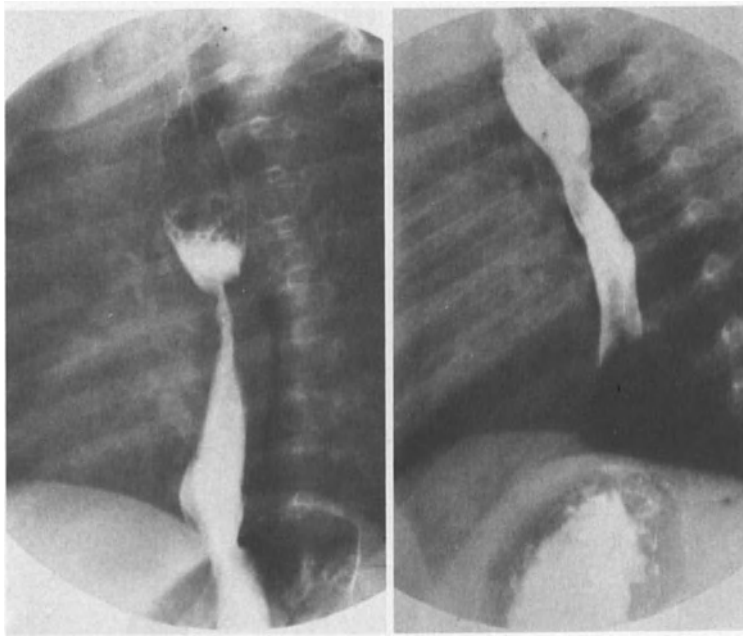
day on, bougienage of the stenosis was done with Rehbein bougies up to Charrière 32. Esophageal manometry on April 28, 1979 showed high cardiac tonus, no gastroesophageal reflux, and well-coordinated swallowing in the distal esophagus, indicating a continuous neuromotoric coordination with synchronous cardia relaxation.

Radiological examination at the age of 14 months showed good esophageal passage without stenosis. The bougienage thread was then withdrawn (Fig. 1f). The child is now 5 years old and entirely free of complaints.

Case 2

G.R., female, born Oct. 17, 1979, 1900 g, 44 cm. Diagnosis was of esophageal atresia without lower fistula, anal atresia with rectovestibular fistula, right-sided duplex kidney, radial aplasia, and Vater's syndrome.

Gastrostomy and colostomy were performed on October 18. The child developed increasing cardiac insufficiency postoperatively. A heart catheter revealed patent ductus arteriosus and a right descending aorta. The child recovered after ductus ligation. Tracheoscopy on November 9 disclosed an upper esophagotracheal fistula. A bougienage thread was inserted endoscopically on December 3 and the upper fistula was closed. During the fourth bougienage (Dec. 12) total atelectasis of the right lung occurred and bronchoscopy showed both bougienage



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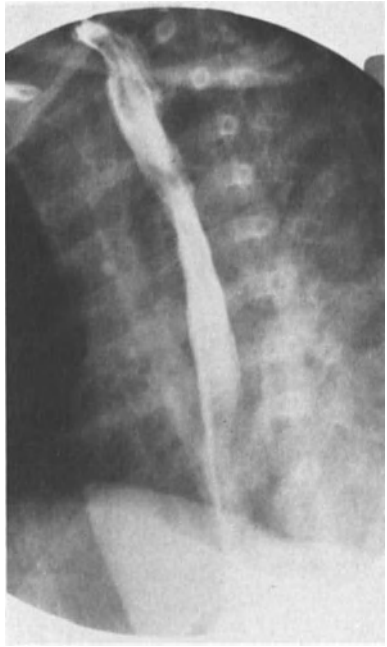


f

Fig. 1d–f

threads running through the right main bronchus; the threads were withdrawn. On December 28, elongation bougienage began according to Howard and Myers (1965); this was repeated ten times until January 11, 1980. A new bougienage thread was installed on Jan. 14 and olive bougienage was restarted and repeated

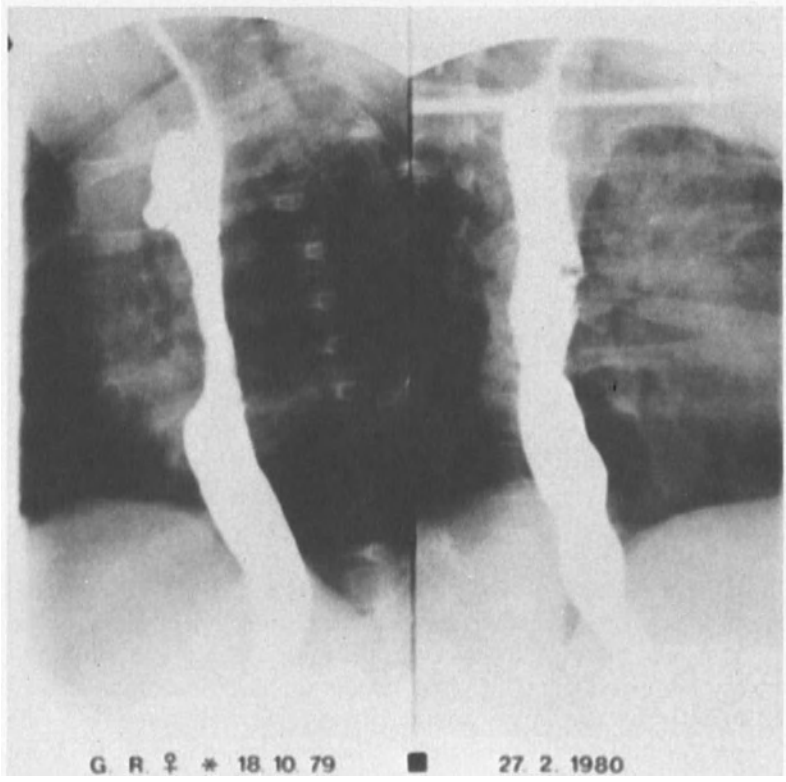


Fig. 2. G.R., born Oct. 17, 1979. Esophageal passage on February 27, 1980. 1 month prior to discharge

eight times. From January 24 on we performed bougienage of the stenosis with Rehbein bougies, 18 times altogether. Esophagoscopy (March 17) revealed stenosis over 3 cm, with granulation tissue. Since the infant swallowed well, she was discharged on March 30 (Fig. 2). She received normal feedings at home and thrived undergoing bougienage in the outpatient department once a week. The last bougienage was performed on June 3, 1980. The child died 2 days later in the ambulance on the way to the clinic. Postmortem examination revealed toxicosis and endocardial fibrosis with lung edema as the cause of death.

Case 3

K.E., female, born Dec. 26, 1981, 1850 g, 43 cm, parents Polish refugees. She was the first child of the father's second wife. His first wife had had two stillbirths and her first child died within 24 h of birth. The diagnosis was of esophageal atresia with esophagotracheal fistula, VSD, multiple malformations of the vertebrae and ribs, right-sided radial dysplasia and thumb hypoplasia, horseshoe kidney, and anal atresia with a rectovestibular fistula.

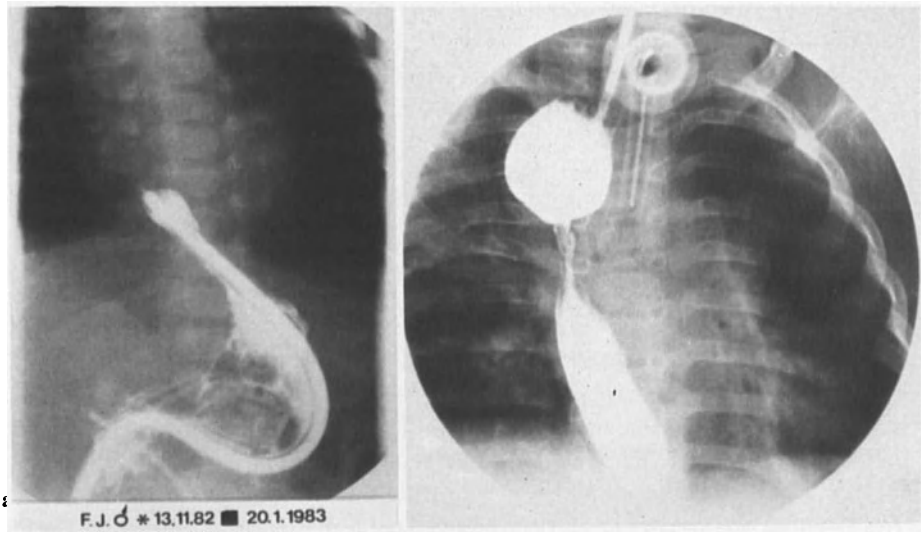
Gastrostomy and anal bougienage were performed on Dec. 27. The child was ventilator dependent and suffered cardiac decompensation on January 8 and renal insufficiency on January 20. A heart catheter (March 11) disclosed a large VSD with left-right shunting and pulmonary hypertension, as well as a small ASD. On March 18 the child was still intubated, but not on the ventilator. She had endoscopic insertion of a bougienage thread on March 20, with olive bougienage from March 23 on, until spontaneous breathing occurred, followed by extubation on March 29. A junction of both pouches was made on April 4, and bougienage of the stenosis was done three times a week. The child thrived. Pulmonary deterioration from May 5 on necessitated feeding via a gastric tube. After an operation for hydrocephalus on May 18 there was further deterioration, and death followed on August 23. Postmortem examination revealed Vater's syndrome, radial dysplasia, thumb hypoplasia, an operated esophageal atresia with stenosis over 1 cm, a VSD with decompensated cor pulmonale, congestive fibrosis of both lungs, multiple brain cysts, and a pelvic kidney.

Case 4

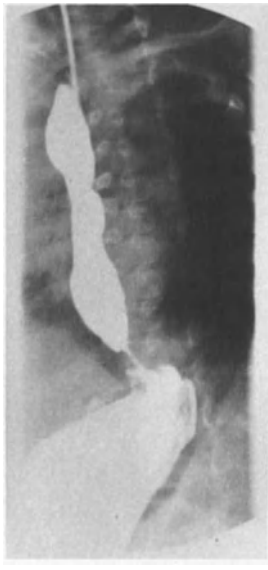
F.J., male, born Nov. 13, 1982, 1250 g, 40 cm. He was put on a ventilator, and underwent gastrostomy on November 15; septicemia developed. He received phototherapy for hyperbilirubinemia; tachycardia was measured up to 200. Extubation was done on December 24, radiography of the distal pouch on Jan. 20 (Fig. 3a). The infant was losing large amounts of potassium and sodium chloride via salivation. We attempted endoscopic thread insertion on January 31, 1983; the thread was running through the left main bronchus and had to be withdrawn. An esophagotracheal fistula was diagnosed in the middle of the trachea. A new sepsis developed. We performed a thoracotomy on February 23, during which an upper esophagotracheal fistula was dissected and closed. Primary anastomosis could not be performed. The child underwent tracheostomy on March 23 for recurrent atelectases which required frequent tracheal suction. We again tried endoscopic thread insertion on April 20 with control tracheoscopy. Olive bougienage was started on April 21 and repeated 13 times, with bougienage of the stenosis from May 5 on. After improvement of his general condition and change to oral feeding, the child was underwent discharged from the hospital with a tracheal cannula on November 25, 1983, and bougienage 1–2 times a week in the outpatient department. Repeated attempts at decannulation failed, obviously owing to esophageal stenosis (Fig. 3b). The stenosis was resected, and esophageal anastomosis performed on May 23, 1984. Radiographic control (June 20) showed good esophageal passage, but gastric dysmotility and massive gastroesophageal reflux. Esophagoscopy revealed extended erosive esophagitis, stage 2–3, which required fundoplication according to Nissen and pyloroplasty, performed on July 7, 1984.

The child is still in the hospital (Fig. 3c) with the tracheal cannula in place.

A fifth patient with long-gap esophageal atresia Vogt-type IIIb, treated by Rehbein's thread-and-olive method, was referred from elsewhere at the age of 1½ years with high-grade esophageal stenosis. The stenosis, which could not be



a b



c

Fig. 3a–c. F.J., born Nov. 13, 1982. **a** Exhibition of lower pouch prior to olive bougienage; **b** esophageal stenosis (March 8, 1984); **c** esophageal passage following resection, fundoplication, and pyloroplasty

treated by bougienage, extended over 3 cm and had to be resected. There were only two small epithelial islets, and epithelium was missing in the rest of the stenosis, which consisted mainly of thick scar tissue, extraordinarily difficult to remove.

Discussion and Conclusions

Remarkably, in all children with lower fistula primary anastomosis could be performed, and only one child operated on elsewhere was treated by Rehbein's tech-

nique. There was an upper esophagotracheal fistula in three of the four patients without lower fistula. Upper fistula was secondarily diagnosed in one case. We have thus concluded that primary tracheobronchoscopy is mandatory in children scheduled for treatment according to Rehbein.

The fact that the main bronchus was perforated during endoscopic thread insertion in two children, which was diagnosed only on the third day in one case, shows on the one hand that this event may be harmless, but on the other that control tracheobronchoscopy is necessary following thread insertion.

It might happen that the thread is torn during olive bougienage; we experienced this once. We now insert a reserve thread prior to first bougienage by means of the thread already in place.

Olive bougienage should be started immediately following thread insertion, and repeated at short intervals of 1–2 days. Therapy can be initiated only if the general condition of the newborn allows it. This was generally the case between the 7th and the 12th week. Comparing the four cases with each other, it becomes evident that the best result was achieved with patient H.J., on whom treatment was begun in the 7th week. Start of bougienage as early as possible seems to be desirable; however, the infant should have reached a weight of at least 2000 g and have overcome septic complications. It turned out to be useful to perform endoscopic thread insertion prior to closure of the esophagotracheal fistula, since otherwise the success of fistula closure would be endangered by subsequent endoscopy.

Our first case shows that excellent results can be obtained by employment of Rehbein's olive technique and that thoracotomy can be avoided by endoscopic thread insertion according to Okmian (Okmian et al. 1975). Normal propulsive esophageal motor function is explained by the fact that both pouches were perforated only by a thin needle, and nervous elements of the esophagus were not damaged by an operation. The result in case 4 cannot be compared with that in case 1, of course. Resection of the esophageal stenosis probably effected gastric dysmotility as well gastroesophageal reflux, Nevertheless, the fact that the child is using his own esophagus may itself be judged as success. Vater's syndrome impaired the prognosis from the beginning for the two infants who died, particularly for case no. 3. In case no. 2, the great distance to our hospital (over 100 km) and the carelessness of the physician who treated the patient were obviously responsible for the fatal outcome. This death and a similar case reported by Kato et al. (1980) should set us to thinking, and we have to conclude that the trend toward as short a hospitalization as possible must not be followed in such cases, at least if adequate aftercare cannot be organized. It must be assumed that bacteria might contaminate the mediastinum, thus endangering the child as long as the wound in the esophagus is granulating. This must be prevented by close supervision and antibiotic therapy if necessary.

Case no. 5 and the cases with type-IIIb atresia reported by Festen (1981) who were treated with Rehbein's olive-and-thread method show that the method obviously does not provide an ideal solution in such instances, since operative resection of an esophageal stenosis might become necessary later on. Here it

seems to be more convenient to enforce primary anastomosis, even under tension, or to render it possible by esophagomyotomy according to Livaditis (Livaditis et al. 1972). An alternative is primary closure of the lower fistula and lower pouch and attachment of the lower to the upper pouch. Later on, a bougienage thread can be inserted endoscopically and olive bougienage employed.

Summary

During the past few years four children with esophageal atresia without lower fistula were treated with the thread-and-olive method according to Rehbein. Our experiences indicate that excellent results can be achieved if the thread is inserted early, endoscopically, and olive bougienage is performed at short intervals. Tracheoscopy should be mandatory prior to the start of treatment, since we observed an upper fistula in three of four cases. Endoscopic thread insertion may lead to perforation of the main bronchus; control tracheobronchoscopy must therefore be done immediately following insertion. Perforation of the main bronchus turned out to be harmless, so the maneuver can be repeated after a few days.

Two of the four infants died of congenital malformations. Therapy was not related to the fatal outcome in either case. One of the survivors has practically normal esophageal motor function with proven propulsive peristalsis. Esophageal stenosis had to be resected in the second survivor; this was followed by massive gastroesophageal reflux with esophagitis and disturbed gastric motility, rendering subsequent fundoplication and pyloroplasty necessary.

Résumé

Durant ces dernières années, 4 enfants avec atrésie œsophagienne sans fistule basse furent traités par fil et olive selon la technique de Rehbein. Notre expérience démontre que d'excellents résultats peuvent être obtenus si le fil est mis en place par voie endoscopique et que le bougirage par olive est réalisé à des intervalles rapprochés. Une trachéoscopie doit être obligatoirement réalisée avant la mise en route du traitement, car nous avons observé une fistule au bout supérieur dans 3 de nos 5 cas. La mise en place endoscopique du fil peut conduire à une perforation d'une bronche majeure. Le contrôle trachéo-bronchoscopique doit donc être pratiqué immédiatement après l'insertion endoscopique du fil. Une perforation d'une bronche majeure cicatrise convenablement et la manœuvre peut être répétée après quelques jours.

Deux de nos 5 enfants moururent de malformations congénitales. Le traitement n'a pas été en relation avec l'issue fatale. Un des suvivants a une motricité œsophagienne pratiquement normale avec un péristaltisme propulsif démontré. La sténose œsophagienne a dû être réséquée chez le 2^{e} survivant, ce qui a été suivi par un reflux gastro-œsophagien massif avec œsophagite et perturbation de la

motricité gastrique nécessitant une fondoplication et une pyloroplastie complémentaires.

Zusammenfassung

In den letzten Jahren wurden 4 Kinder mit Ösophagusatresie ohne untere Fistel mit der Olivenmethode nach Rehbein behandelt. Die Erfahrungen zeigen, daß es möglich ist, damit ein ausgezeichnetes Resultat zu erzielen, wenn es gelingt, frühzeitig (endoskopisch) den Faden zu legen und die Olivenbougierungen in kurzfristigen Abständen durchzuführen. Da bei 3 von 4 Fällen eine obere Fistel beobachtet wurde, sollte vor Beginn der Behandlung unbedingt eine Tracheoskopie durchgeführt werden. Die endoskopische Fadenlegung kann zur Perforation eines Hauptbronchus führen, daher muß unmittelbar nach der Fadenlegung eine Tracheobronchoskopie durchgeführt werden. Die Punktion des Bronchus hat sich als harmlos herausgestellt, das Manöver kann daher nach wenigen Tagen wiederholt werden.

Von 4 Kindern verstarben 2 an ihren kongenitalen Mißbildungen während der Behandlung. Die Behandlung selbst war in beiden Fällen am letalen Ausgang ursächlich nicht beteiligt. Von den Überlebenden hat ein Kind eine praktisch normale Ösophagusfunktion mit nachweisbarer, propulsiver Peristaltik. Bei dem zweiten Kind mußte die Ösophagusstenose reseziert werden. Nach dieser Operation kam es zu einem massiven gastroösophagealen Reflux mit Ösophagitis und einem Antrumdysmotilitätssyndrom. Es war daher im weiteren Verlauf noch eine Fundoplikation mit Pyloroplastik notwendig.

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Incidence of Gastroesophageal Reflux Following Repair of Esophageal Atresia

A. Koch[1], S. Rohr[1], J. Plaschkes[1], and M. Bettex[1]

With the increase in attention paid to the problem of gastroesophageal reflux (GER) there is a rise in the number of publications emphasizing the predisposition of repaired esophageal atresia to GER and its aggressive prevention and therapy (Ashcraft et al. 1977; Fonkalsrud 1979; Pieretti et al. 1974).

To clarify particularly controversial statements on the incidence of GER following repaired esophageal atresia and the necessity of treatment, different groups of patients treated at different times were investigated retro- and prospectively by different methods.

Patients

Group I consisted of 27 patients, aged 16–23 years and operated on elsewhere from 1951 to 1958 who were followed up clinically, radiologically, and manometrically, and the majority of them (23) also endoscopically (Table 1).

Group II consisted of 28 patients, aged 4–14 years, who were operated on at our hospital and prospectively followed up clinically and radiologically at intervals of 3–6 months initially, later at intervals of 12–24 months. When discrepancies between clinical and radiological findings were seen, esophagoscopy was employed (12/28). For the same reason manometry – available only since 1979 – was performed seven times.

Group III was 12 patients who were also prospectively observed for 1.5–5 years. In addition to the previously mentioned diagnostic procedures 24-h pH monitoring was used in this group.

Table 1. Retro- or prospectively observed groups after repair of esophageal atresia

Group	Patients *(n)*	Period of primary repair	Follow-up (years)
I	27	1951–1958	16 –23[a]
II	28	1970–1979	4 –14
III	12	1979–1982	1.5– 5

[a] Koch et al. (1976)

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Table 2. Local complications after primary repair of esophageal atresia

Complications	Group		
	I	II	III
Recurrent fistula	1/27	3/28	1/12
Anastomotic stenosis requiring	14/27	12/28	3/12
– Dilatation >2×	14[a]	8[b]	3
– Resection	–	4	–

[a] 5× gap >3 cm
[b] 1× under minimal tension
1× under considerable tension

Anastomotic complications (Table 2) influencing the further course arose predominantly from fistula relapse and stenosis. Among the oldest group (I) an entirely asymptomatic recurrent fistula was diagnosed radiologically in a 17-year-old patient. In contrast, recurrent fistulas were diagnosed and corrected four times within the first year of life in both younger groups. Stenoses resistant to long-term bougienage required resection four times in the second group exclusively. In cases which required bougienage for stenosis the primary gap had been described in five cases as more than 3 cm, and tension to the anastomosis was "low" and "marked" respectively in two others.

Methods

On control investigations GER was looked for and judged on the basis of frequency and extent of vomiting as well as its esophageal, pulmonary, and general complications; these were registered by anamnesis and actual symptoms. In adolescents and adults retrosternal burning or occasional rumination were taken as hints of GER.

Radiological investigation was done as an upper gastrointestinal series. In group I provocation measures were done by abdominal pressure, in groups II and III by the water-siphon test. Only repeatedly proven supradiaphragmatic displacement of parts of the stomach was judged as hiatal hernia.

Flexible esophagoscopes (Olympus GIFK) were used exclusively in group I and rigid ones in groups II and III (Storz, 9 mm). Inflammatory changes were classified in three degrees, ranging from erythema over localized erosions to generally confluent longitudinal and circular erosions.

Manometry was performed as 3-point measurement under permanent perfusion (Koch and Rüggeberg 1978). We used probes with side holes 5 cm apart from each other in group I and 3 cm in groups II and III. Resting pressure and swallow reflexes of the lower esophageal sphincter (LES) were judged. Simul-

Table 3. Characteristics of pathological GER

1. Presenting symptoms	– Regurgitation of gastric contents $>3\times/24$ h – Failure to thrive – Documented repeated aspiration – Epigastric or retrosternal pain
2. Barium swallow	– Spontaneous and repeated return of contrast-medium into the lower esophagus during a single investigation – "Fixed" hiatal hernia
3. Esophagoscopy	– Esophagitis (erythema, erosion) and its sequelae (stenosis, endobrachyesophagus)
4. Esophageal manometry	– Resting pressure LES ≤ 12 mm Hg – Common cavity phenomenon spontaneously or on abdominal compression
5. Extended pH monitoring	– Total reflux duration $>4\%$ – Reflux duration during sleep $>1.5\%$ of measuring time

taneously, we looked for spontaneous and (by abdominal pressure) provoked conduction of the fundus pressure wave into the esophagus. In group I hormonal stimulation of the LES was investigated by intravenous administration of pentagastrin (0.6 g/kg) (Koch et al. 1976).

For 24-h pH monitoring we used a combination electrode especially adapted to infants (Koch and Gass 1981). All reflux episodes defined as pH decrease to below 4 over at least 30 s were recorded absolutely and in percent of total monitoring time. The second essential parameter was relative reflux duration during sleep.

All findings characteristic for pathologic reflux are summarized in Table 3.

Results

According to the parameters mentioned (Table 4), seven patients of group I had GER-positive findings: two endoscopically, as low-grade esophagitis and a short endobrachyesophagus respectively; six manometrically as esophageal conduction of fundus pressure waves provoked by abdominal pressure (common cavity phenomenon) in normotone- and pentagastrin-stimulated resting pressure of the LES. Only one of the patients with endoscopic findings also had manometrically determined sphincter insufficiency. All positive findings were related to patients who presented neither anamnestically nor radiologically with signs of GER.

In group II, diagnosis of GER was based mainly on clinical and radiological findings in 21 patients. GER was associated with endoscopically verified low- to medium-grade esophagitis in five cases and with endobrachyesophagus in another case. Sphincteric hypotension was proven manometrically in two cases. The majority of GER-positive results were related to those ten patients subsequently

Table 4. Positive findings for pathological GER after primary repair of esophageal atresia in the various age groups

Diagnosis	Group		
	I	II	III
Clinical presentation	0/27	9/28[a]	4/12
Barium swallow	0/27	16/28[b]	8/12
Esophagoscopy	2/23	6/28[c]	1/12[d]
Manometry	6/27	2/ 7	0/12
24-h pH monitoring	–	–	3/12[e]
Found in	7/27	21/28	9/12

[a] Five operated
[b] Eight operated
[c] Three operated
[d] One operated
[e] None operated

Table 5. Number and combination of pathological reflux findings after primary repair of esophageal atresia in various age groups

No. of pathological findings	Group		
	I	II	III
1	6 (5 × mano) (1 x endo)	11 (7 × rx) (3 × clin) (1 × mano)	3 (3 × rx)
2	1 (1 × endo + mano)	8 (4 × rx + clin) (3 × rx + endo) (1 × clin + endo)	5 (2 × rx + clin) (2 × rx + pH-metr) (1 × clin + pH-metr)
3	–	2 (1 × rx + clin + endo) (1 × rx + endo + mano)	1 (1 × rx + clin + endo)
Total	7/27	21/28	9/12

clin = clinical; rx = radiological; endo = endoscopic; mano = manometrical; pH-metr = pH monitoring

operated on and were found during the first year of life, with the exception of one 2-year-old boy with severe cerebral palsy.

In the group of youngest patients (group III) GER-positive findings were obtained, nearly without exception, during the first 3–6 weeks of life. In only one case was a diagnosis of medium-grade esophagitis made at the end of the 2nd year, despite intermittent vomiting and radiologically proven spontaneous reflux

during control investigation performed elsewhere. Clinically and radiologically pathological results were found in eight infants. In the patient with endoscopically positive findings pH monitoring was negative, whereas three positive pH monitoring results were found, in a clinically and radiologically normal infant and in two children with radiologically and clinically GER-positive signs. Radiological diagnosis was not possible in four infants in whom filling of the stomach with contrast medium was insufficient. Comparison of the number and kind of positive GER findings showed that in the oldest group, 75% of the patients had no positive signs, whereas in both younger groups, 75% presented with positive signs (Table 5).

Single and double GER-positive findings were most frequently encountered; triple-positive findings were the exception, and not found in the oldest patients (group I) at all. Most frequently, pathological findings were made by radiological examination, followed by clinical examination.

Reflux Therapy and Course

Therapeutic measures seemed to be unnecessary in group I until the control investigation mentioned, and later since the patients were free of complaints. Conservative treatment predominated in groups II and III, but operative measures were relatively frequently employed. Indication for operation was based mainly on repeated positive radiological and clinical findings (Table 6).

Only in one case of a resected anastomotic stenosis, where brachyesophagus and partial thoracic displacement of the stomach were found, did radiological findings alone seem sufficient for antireflux surgery. Endoscopically proven low- to medium-grade esophagitis in radiologically and clinically persistent findings was decisive for surgery in five cases. With the exception of two cases of severe cerebral palsy and delayed endoscopy, all operative procedures were performed during the first year of life, particularly during the first 4 months. Nissen's fundoplication was used exclusively. More than half of the patients operated on in this

Table 6. Indications for antireflux operation after repair of esophageal atresia

Indication	Group		
	I	II	III
Radiological/clinical	–	5	–
Radiological	–	1	–
Endoscopic/radiological	–	2	–
Endoscopic/clinical	–	2[a]	1[a]
Total	0/27	10/28	1/12

[a] Not operated until 2 years of age: 2 × severe cerebral palsy
1 × controlled elsewhere

Table 7. Antireflux operations related to early anastomotic complications

Complications	*n*	Antireflux operation		
		Group I	Group II	Group III
Recurrent fistula	5	–	2	1
Anastomotic stenosis requiring	29	–	3	–
– Dilatations >2×	25	–	–	–
– Resection	4	–	3	–

Table 8. Evolution of prospectively controlled GER under conservative management after repair of esophageal atresia

Group	*n*	No. of normalizations (after)	No. of improvements (after)
II	11	6 (1 year)	–
		5 (4–9 years)	–
III	8	4 (1 year)	2 (1 year)
		2 (2 years)	–

way (Table 7) had developed anastomotic complications such as fistula recurrence and early stenosis, following esophageal repair. In contrast, there was no manifestation of GER requiring therapy in two cases following closure of fistula and in another after resection of stenosis. Under conservative reflux therapy (Table 8) clinical and radiological findings normalized mainly during the first 12 months, whereas delayed normalization was due exclusively to changing radiological GER. Two cases showed clear improvement of clinical and radiological signs of a low-grade persistent GER at the time of follow-up.

Discussion

An increased tendency toward GER following esophageal repair is undisputed today. Causes for this are delayed emptying of the stomach, cranial displacement of the esophagogastric junction diminishing antireflux function, and decrease in or even loss of clearance ability in the hypo- or aperistaltic esophagus.

The subject of permanent controversy, however, is the incidence of GER and its consequences following esophageal repair. The considerable divergence of incidence rates published (Table 9) was also found in our groups of patients

Table 9. Published incidence of GER after repair of esophageal atresia (TEF) related to different diagnostic methods

Publication	GER/ TEF	Diagnostic procedure
Laks et al. (1972)	0/ 15	rx
Johnson et al. (1977)	5/ 55	rx, mano, ART
Orringer et al. (1977)	13/ 22	rx, mano, ART
Fonkalsrud (1979)	9/ 14	rx
Jolley et al. (1980)	12/ 25	pH-metr, scinti
Gauthier et al. (1980)	58/113	rx

rx = radiology; mano = manometry; ART = acid reflux test; pH-metr = pH monitoring; scinti = scintiscanning; TEF = tracheoesophageal fistula

followed up. As it becomes clear from the publications mentioned, the most important cause for discrepancy is the lack of a uniform definition of GER based on the method of investigation. All methods of directly diagnosing GER, such as contrast-medium passage, scintiscanning, and long-term pH monitoring, tend to overestimate GER unless pathological GER is clearly defined by comparison with control groups of the same age and by prospective investigations. During early infancy reflux can be relatively easily induced by sufficient filling of the stomach with contrast medium, whereas even strong provocation maneuvers remain without effect in adolescents and adults (group I). Thus, it is not surprising that most authors who did their follow-ups relatively early report positive radiological GER findings in the majority of cases (Fonkalsrud 1979; Gauthier et al. 1980) as we found in both of our younger groups. GER diagnosed this way can be judged correctly only by a combination of clinical and endoscopic findings. The same is true for pH monitoring: only determination of pathological reflux duration regarding a sharply defined age group with its physiological characteristics may avoid false incidence rates, which are particularly found where unphysiologic acid substances were used (Jolley et al. 1980; Orringer et al. 1977). The rare correlation between pH monitoring findings and reflux symptoms in these publications, as well as in group III of our study, emphasizes the significance of clinical signs and endoscopy as far as necessity for and kind of treatment are concerned.

This is highly valid for the so-called pulmonary complications of GER, which cannot be causally explained even with the most intense supervision during pH monitoring (Koch et al. 1985). The urgency of operative treatment can be based only on repeatedly registered aspiration requiring reanimation, but neither on number and duration of reflux episodes nor on radiologially and scintigraphically observed passage of gastric contents into the airways. Those methods which disclose only partial aspects of reflux pathogenesis, such as manometry, or only esophageal complications, such as esophagoscopy, carry a different weight than do methods of direct GER diagnosis. It follows that simple addition of positive

findings, as was done for GER classification by several authors (Parker et al. 1979), cannot be sufficient.

The second essential cause of discrepancy of incidence rates, tightly linked to the first, regards the patient's age when the diagnosis was made. This factor was not considered in the publications presenting high incidence rates (Orringer et al. 1977; Parker et al. 1979). However, the majority of authors confirm our experience, that need for therapy in general and for surgery in particular is decided on almost exclusively during the first year of life (Ashcraft et al. 1977; Gauthier et al. 1980; Jolley et al. 1980). The infrequent follow-ups during early infancy (Gauthier et al. 1980) as well as retrospective studies of predominantly asymptomatic adults (Burgess et al. 1968; Laks et al. 1972; Orringer et al. 1977) confirm our observation that GER diagnosed beyond the first year of life is seldom of clinical significance.

Our follow-up over 1.5–14 years of 19 conservatively treated patients, the majority of whom showed normalization within 1 year with only exceptions after several years, disproves the assumption of some authors that GER manifested following esophageal repair does not improve (Ashcraft et al. 1977; Fonkalsrud 1979; Orringer et al. 1977).

Consistent follow-up until termination of growth is therefore necessary to confirm our experience or to disclose exceptionally different courses.

Besides diagnostic problems, incidence of GER seems to be particularly dependent on intra- and early postoperative anastomotic complications. Only a few authors found a direct relation between width of gap, extent of mobilization, and anastomotic tension and GER documented by pH monitoring (Orringer et al. 1977). This was only exceptionally possible in our patients, since anatomical conditions were insufficiently documented. However, our cases with fistula relapse and stenosis requiring resection indicate a strong tendency of anastomotic complications to manifest as GER. Therefore, those cases where antireflux function of the esophagogastric junction might be impaired by esophageal shortening give occasion for a more intense follow-up.

We did not observe anastomotic stenosis caused by GER in our patients, as described by Pieretti et al. (1974) in an indefinite number of cases.

Conclusions

Factors affecting the incidence of GER following primary repair of esophageal atresia are:

1. The method used to diagnose reflux
2. The definition of reflux
3. Patient's age at examination
4. Anastomotic complications

GER can be diagnosed only by considering both actual symptoms and endoscopically documented complications. Certain anastomotic complications necessitating another operation, add to the risk of developing reflux. These cases in particular require long-term follow-up. While therapy does not differ from the

general principles of treatment for reflux (Koch et al. 1982), there may be problems with relative indications in this kind of underlying disease. Thus, it is our opinion that it is more important to keep in mind the particular tendency of GER to appear following esophageal repair and to make a proper diagnosis, taking the specific aspects of the individual methods into consideration, than it is to heed incidence rates.

Summary

Gastroesophageal reflux (GER) was looked for retro- and prospectively by various diagnostic methods in 67 patients, subdivided into three groups according to age at time of investigation.

Pathological reflux was found in only one-quarter of the patients aged 16–23 years, but in three-quarters of both younger groups followed up for 1.5–14 years so far. Whereas pathological findings were most frequently disclosed by manometry in the older patients, radiological findings were most often decisive in younger patients. Following closure of recurrent tracheoesophageal fistulas and resection of anastomotic stenoses, eight patients developed reflux complications during the first months of life; these were so severe that antireflux plasty became unavoidable.

Factors influencing the incidence of GER following esophageal repair were: the definition of GER based on the diagnostic method employed, the patient's age at the time of diagnosis, and anastomotic complications requiring a second operation. The essential time for treatment was the first year of life.

Résumé

Les caractéristiques du reflux gastro-œsophagien furent précisées par études rétro- et prospectives par les différentes méthodes de diagnostic chez 67 patients subdivisés en 3 groupes en fonction de l'âge au moment des investigations.

Un reflux pathologique fut trouvé chez seulement un quart des patients âgés de 16 à 23 ans; dans les 2 groupes couvrant la période de 1,5 à 14 ans, ce reflux fut observé dans les trois quarts des cas. L'anomalie pathologique la plus fréquente fut d'ordre manométrique chez les patients les plus âgés et d'ordre radiologique chez les plus jeunes. Après fermeture des fistules trachéo-œsophagiennes récidivantes et résection anastomose des sténoses effectuées pendant les premiers mois de la vie, 8 patients développèrent un reflux tellement important qu'une opération antireflux devint nécessaire.

Les éléments influençant l'incidence du reflux gastro-œsophagien après réparation de l'œsophage ont été: relations entre la méthode diagnostique employée, l'âge au moment du diagnostic et les complications de l'anastomose nécessitant une autre opération.

Le meilleur moment pour le traitement a été la première année de la vie.

Zusammenfassung

Bei 67 Patienten, die entsprechend ihrem Alter zum Zeitpunkt der Untersuchung in 3 Gruppen unterteilt waren, wurde mit verschiedenen diagnostischen Verfahren retro- bzw. prospektiv ein für einen gastroösophagealen Reflux (GOR) charakteristischer Befund gesucht.

In der Gruppe der 16- bis 23jährigen Patienten fand sich ein pathologischer Reflux nur bei 25%, in den beiden Gruppen der bislang über 1,5- bis 14jährigen beobachteten hingegen bei 75%. Häufigster pathologischer Befund war unter den älteren Patienten der manometrische, unter den jüngeren der radiologische Befund. Bei 8 Patienten stellten sich nach Verschluß einer rezidivierten tracheoösophagealen Fistel bzw. Resektion einer Anastomosenstenose bereits in den ersten Lebensmonaten so ausgeprägte Refluxfolgen ein, daß eine Antirefluxoperation für unumgänglich erachtet wurde.

Somit waren die Faktoren mit dem wesentlichsten Einfluß auf die Refluxinzidenz nach Anastomosierung einer Ösophagusatresie: die Refluxdefinition in Abhängigkeit vom angewandten diagnostischen Verfahren, das Lebensalter zur Zeit der Refluxdiagnostik und Anastomosenkomplikationen, die zu einem Zweiteingriff geführt hatten. Als Zeitraum, der für die Therapie am entscheidensten war, erwies sich das 1. Lebensjahr.

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Pressure-Induced Growth (PIG) of Atretic Esophagus: A Contigent Management for High-Risk Esophageal Atresia

W.-M. Pieper[1], S. Hofmann-von Kap-herr[2], and D. nii-Amon-Kotei[1]

The management of uncomplicated esophageal atresia is a straightforward procedure consisting of thoracotomy with anastomosis of the atretic segments and ligature of any tracheoesophageal fistula present. However, besides the rareness of isolated esophageal atresia, our observations in the past few years show increasing incidence of the anomaly in association with premature births and other malformations which are usually incompatible with life. Aspiration pneumonia resulting from reflux of gastric content through a lower tracheoesophageal fistula, rather than the overflow of saliva from an upper atretic pouch, also threatens the life of an otherwise healthy neonate with esophageal atresia.

A number of procedures designed as parts of a staged management do not, however, always meet their intended aim of safeguarding against the high risk during the management of complicated esophageal atresia, especially of those with aspiration pneumonia. Whereas Koop's (Koop and Hamilton 1965) fistula ligature and gastrostomy does not avoid the extensive and additionally traumatizing thoracotomy, Randolph's (Randolph et al. 1968) gastric division can have many complications.

The relatively minor abdominal cardia banding Leininger (1972) can leak, producing reflux, or tissue necrosis can result from overtightening of the Silastic band.

Hofmann (1975, 1976) therefore introduced balloon catheter blockage, which allows for temporary feeding by a first catheter inserted into the stomach via gastrostomy, and for the blocking of a lower tracheoesophageal fistula by means of a second catheter, also inserted through the gastrostoma, placed in the lower atretic esophageal pouch, and secured in position by a Silastic band around the cardia. This has the aim of preserving the life of the patient until such time that final and definite anastomosis can be performed. The balloons of both catheters are inflated to half their capacity to prevent pressure necrosis. The reflux of gastric content into the esophagus, and thereby aspiration through the tracheoesophageal fistula is prevented by the higher pressure of the artificial respiration, transmitted through the fistula. As a side-effect an enlargement of the lower esophageal pouch was observed, and has been found in animal experiments to be a gross hypertrophy of the esophageal wall, with a constant ratio of cellular to intercellular tissue components. We have termed the procedure "pressure-induced growth" (PIG).

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Nine children have undergone this procedure; five were high-risk cases with multiple anomalies, and eight who presented with long-gap atresia also underwent the Rehbein method of elongation. Five children died of their multiple malformations and one because of septicemia. Three children, however, now aged 8 months, 5 years, and 8 years, have a good esophageal passage after the delayed and final anastomosis. This demonstrates the effectiveness of the procedure, which, however, proves advantageous only when applied in high-risk cases under artificial respiration.

The prevention of aspiration pneumonia, induction of growth of atretic segments, and above all the application of the method as the first part of a staged management of esophageal atresia in high-risk cases are the advantages of PIG.

Summary

A method is described by which a balloon catheter is inserted into the lower pouch, avoiding aspiration pneumonia due to overflow of gastric contents via a lower tracheoesophageal fistula in high-risk cases of esophageal atresia. As a side-effect there is an enlargement of the lower pouch, termed by the authors "pressure-induced growth". The prevention of aspiration pneumonia, induction of growth of the atretic segment, and the application of this method as the first part of a staged management of esophageal atresia in high-risk cases are the advantages.

Résumé

Dans les cas d'atrésie œsophagienne avec fistule trachéo-œsophagienne basse à hauts risques, les auteurs décrivent une méthode ultilisant un cathéter à ballonnet et qui peut être mis en place dans le cul-de-sac œsophagien inférieur pour éviter les pneumonies par aspiration du liquide gastrique. Comme effect annexe, ce système amène un élargissement de la poche inférieure stimulant la croissance. Les auteurs insistent sur les avantages de cette méthode dans la prévention des pneumonies par aspiration, dans l'induction d'une croissance stimulée du segment atrétique, de telle sorte qu'ils recommandent son application comme premier temps du traitement différé d'une atrésie œsophagienne.

Zusammenfassung

Es wird eine Methode beschrieben, die unter Verwendung eines Ballonkatheters, der in das untere Segment eingeführt ist, bei Fällen von Ösophagusatresie mit hohem Risiko eine Aspirationspneumonie durch Übertritt von Mageninhalt über die untere Fistel verhindert. Als Nebeneffekt kommt es zu einer Größenzunahme des unteren Segments, von den Autoren „druckinduziertes Wachstum" bezeichnet. Die Verhinderung einer Aspirationspneumonie, Wachstumsinduktion des

atretischen Segments und die Anwendung dieser Methode als erster Schritt einer mehrstufigen Behandlung bei Ösophagusatresie in Risikofällen sind die Vorteile der Methode.

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Dacron-Patch Aortopexy

L. Spitz[1]

The classical technique involved in performing an aortopexy consists of placing the sutures for traction to the posterior surface of the sternum through the wall of the aorta. These sutures are deliberately passed through the adventitial and medial layers of the aortic wall in an attempt to avoid entering the intimal layer. The traction forces exerted on the aorta in anchoring it to the back of the sternum are therefore distributed over a relatively small surface area. The sutures may cut out and the likelihood of long-term permanent damage to the wall of the aorta owing to aneurysmal degeneration cannot be completely dismissed. The dacron-patch aortopexy was devised to minimize trauma to the wall of the aorta by:

1. Attaching the patch to the aortic wall by means of a number of fine sutures
2. Passing the heavy sutures for the aortopexy through the dacron patch

Traction on the aorta is evenly distributed over a large area rather than concentrated on three or four specific points.

Technique

The operative approach is via an anterior thoracotomy through the bed of the unresected left third rib. The left phrenic nerve is identified and carefully retracted. The left lobe of the thymus is mobilized from the anterior surface of the ascending aorta and gently retracted medially. The anterior surface of the ascending aorta and arch of the aorta is carefully exposed.

A dacron patch approximately 1.5 cm × 0.5 cm is attached to the anterior surface of the ascending aorta and arch of the aorta using either a continuous or an interrupted 5.0 prolene suture (Fig. 1).

Three or four 2.0 prolene sutures are now placed through the central part of the dacron patch. These sutures traverse the manubrium sterni to emerge on the anterior surface of the sternum. When all three or four sutures are in position they are tied in sequence, while an assistant gently depresses the sternum towards the aorta (Fig. 2).

On completion of this maneuver the dacron patch should lie snugly, immediately adjacent to the posterior surface of the manubrium sterni.

This procedure has been carried out in eight patients with tracheomalacia. All eight have responded dramatically with resolution of the stridor, the apneic

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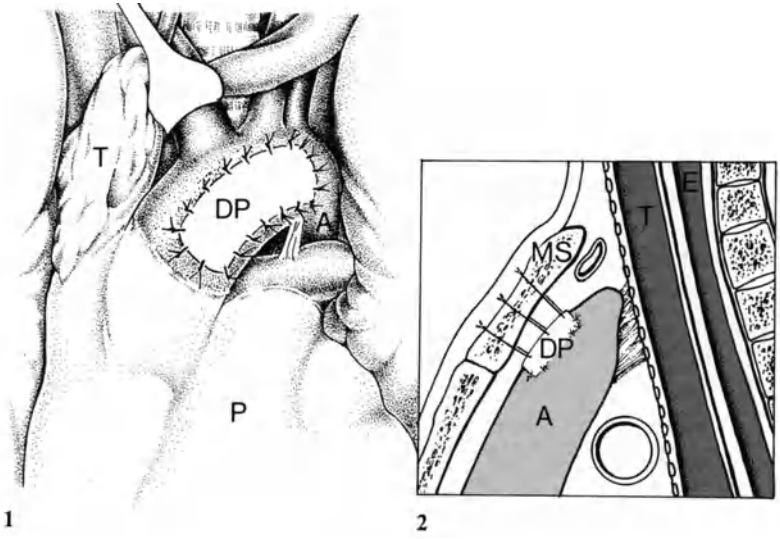

Fig. 1. Dacron patch attached to the anterior wall of the ascending aorta and arch of the aorta. *T,* thymus; *DP,* dacron patch; *A,* aorta; *P,* pericardium

Fig. 2. Completed aortopexy. *MS,* manubrium sterni; *A,* aorta; *T,* trachea; *E,* esophagus; *DP,* dacron patch

attacks, and the respiratory infections. The only complication was the development of a mediastinal hematoma in one infant, which presumably originated from the mobilization of the left lobe of the thymic gland.

Summary

A new technique for aortopexy is described, which uses a dacron patch. The advantages of this procedure as compared with the classical technique are minimized trauma to the aortic wall and evenly distributed traction on the aorta over a large area. This technique was carried out successfully in eight patients with tracheomalacia.

Résumé

Une nouvelle technique d'aortopexie est décrite qui utilise un patch en dacron. En comparaison avec les techniques classiques, les avantages de ce procédé sont de minimiser le traumatisme sur la paroi aortique et de distribuer la traction sur l'aorte sur une large surface. Cette technique a été utilisée avec succès chez 8 patients porteurs de trachéomalacies.

Zusammenfassung

Es wird eine neue Technik der Aortopexie unter Verwendung eines Dacron-patch beschrieben. Die Vorteile dieser Methode gegenüber der klassischen Aortopexie sind: minimale Traumatisierung der Aortenwand sowie gleichmäßiger, über eine größere Fläche verteilter Zug an der Aorta. Diese Methode wurde erfolgreich an 8 Patienten mit Tracheomalazie angewendet.

Reconstruction of the Thoracic Esophagus Using Autotransplanted Small Intestine – An Experimental Study in the Piglet

G. MALMFORS[1] and L. OKMIAN[1]

Several methods have been proposed to correct esophageal defects with bowel substitutes in the treatment of long-gap esophageal atresia. The most commonly used methods are the specific colonic interposition procedures named after Waterston and Sherman. A common disadvantage of these methods is that a huge amount of bowel must be used to bridge a rather small distance between the esophageal endings and all esophageal tissue available cannot be used for its original purpose.

The experimental study here discussed aimed at a new method for bridging an esophageal defect with a vascularized jejunal autotransplant (Malmfors et al. 1981). Free jejunal autotransplants have been successfully used in treating adult patients with esophageal defects after resection of pharyngeal cancers (Seidenberg et al. 1959). A few attempts have been made experimentally to bridge intrathoracic esophageal defects with free bowel autotransplants. With one exception (Meeuwis et al. 1980), these attempts have been failures.

Earlier investigators have found one of the significant problems to be the bending of the relatively long vascular pedicle. The new concept in our study was to avoid this problem by anastomosing the transplant vessels end-to-side, directly to the great intrathoracic vessels, thereby reducing the length of the vascular pedicle.

Figure 1 is a sketch of the operative procedure used, with a jejunal autotransplant interposed between the esophageal endings and with microvascular anastomoses between the transplant artery and aorta and between the transplant vein and the azygos vein.

Method

The study was performed with piglets weighing approximately 6 kg. The reasons for using piglets are that they are extremely fast growing and therefore especially suitable for studying pediatric surgical questions and that they are relatively cheap and easily available. The operations were performed with the piglets under ventilator anesthesia and with muscle relaxation. The operations were started in the abdomen, where a suitable jejunal segment was chosen and extirpated with its vascular pedicle. The transplant artery was catheterized with a 1-mm silicon

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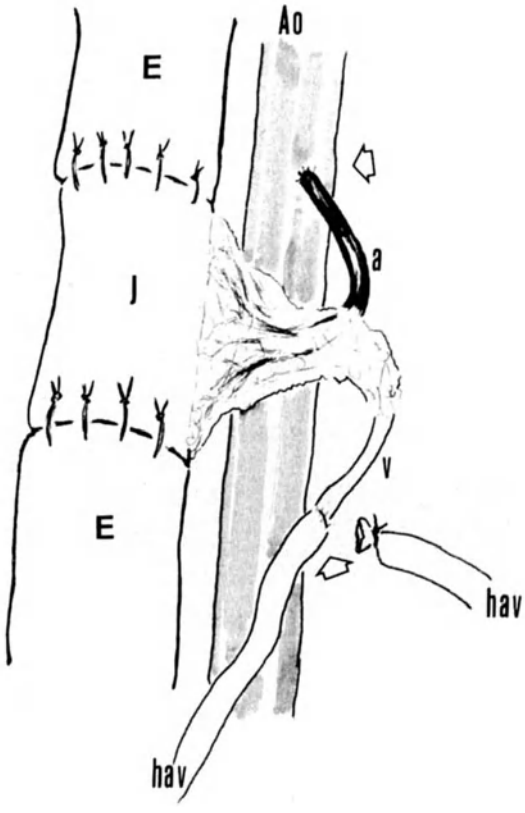

Fig. 1. Schematic representation of operative procedure. *E,* esophagus; *J,* jejunal autotransplant; *a,* transplant artery; *Ao,* aorta; *v,* transplant vein; *hav,* hemiazygos vein

rubber catheter, and the transplant was perfused with a cool heparinized dextran-electrolyte solution (Fig. 2).

After restoration of intestinal continuity the abdomen was closed, and a left-sided thoracotomy was performed in the fifth intercostal space and the esophagus, the aorta and the hemiazygos vein were dissected free. The aorta was partially clamped before a suitable hole was made with specially designed scissors which cut a conical hole (Fig. 3). This facilitated an anastomosis between the tiny transplant artery and the relatively thick aortic wall (Malmfors 1981). The anastomosis was sewn under magnification with a 9/0 or 10/0 ethilon running suture. The venous anastomosis was performed between the transplant vein and the hemiazygos vein either end-to-side or end-to-end, depending on the width of the hemiazygos vein.

After the transplant was revascularized a few centimeters of esophageal tissue was resected and the autotransplant was adapted to suit the esophageal defect. The anastomoses between the esophagus and the autotransplant were performed end-to-end in one layer with interrupted silk stitches. The thoracotomy was closed without drains. Some piglets had a transanastomotic tube while others had a gastrostoma.

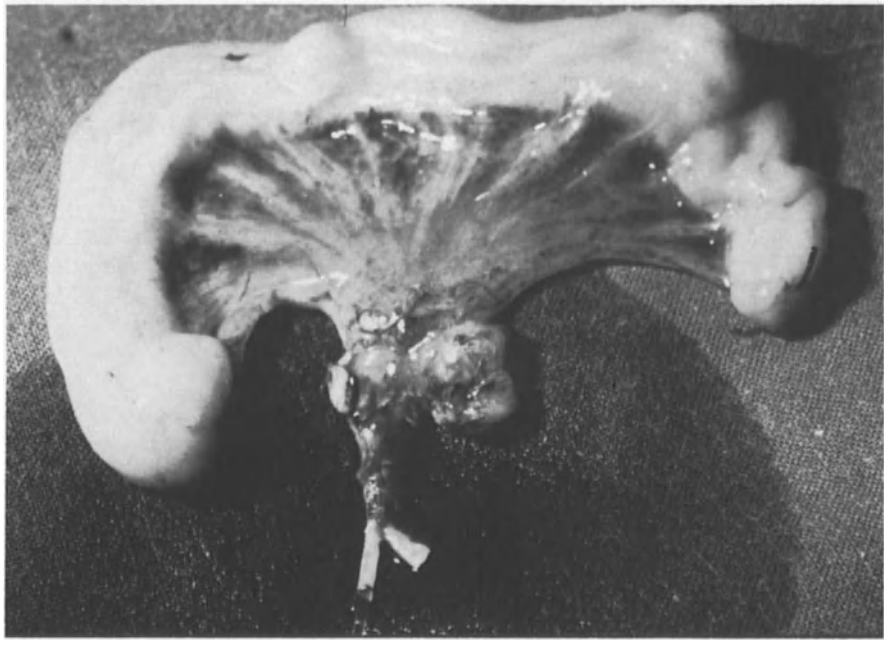

Fig. 2. Autotransplant during perfusion via 1-mm catheter in the artery

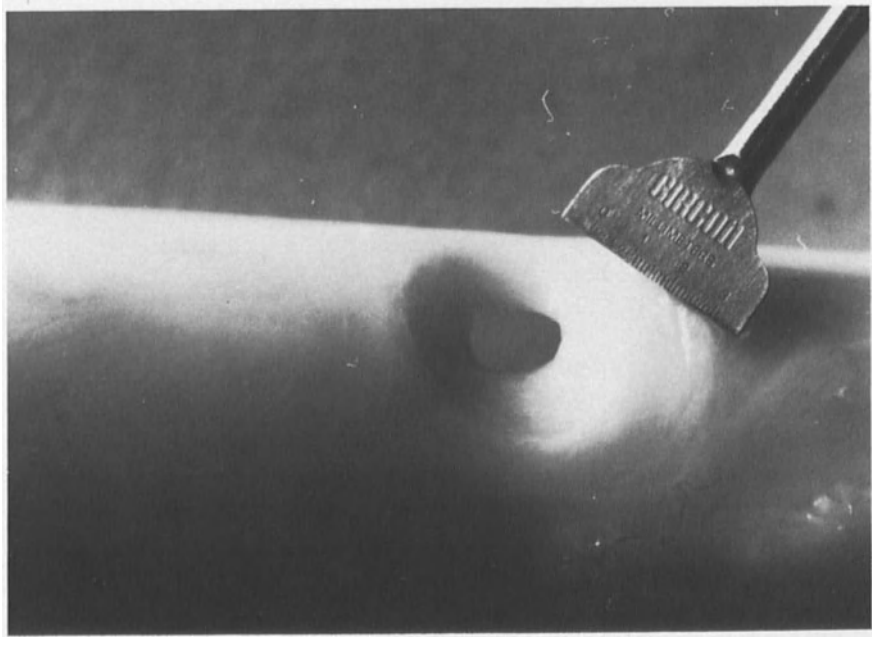

Fig. 3. Conical hole in aorta

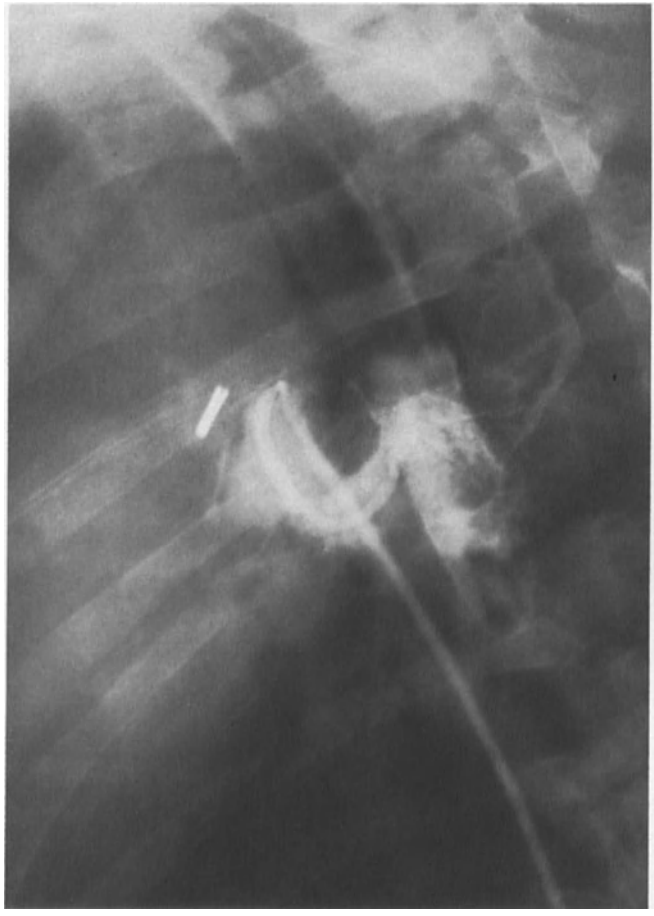

Fig. 4. Angiographic view of jejunal autotransplant interposed as esophageal substitute; catheter in aorta

Results

The operative procedure tested was difficult and very demanding. In the beginning of the series a number of operations had to be interrupted as the revascularization was unsuccessful. However, in two-thirds of the operations the revascularization of the transplant was successful; that is, there were pulsations in the transplant and there were no signs of venous congestion. Even when the revascularization was successful, several piglets died of various postoperative complications such as hypothermia, acidosis, postoperative bleeding, intestinal obstruction, and transplant necrosis leading to mediastinitis.

Figure 4 is a selective angiograph of the autotransplant in one case. The catheter is in the aorta at the site of the microvascular anastomosis. When the piglet

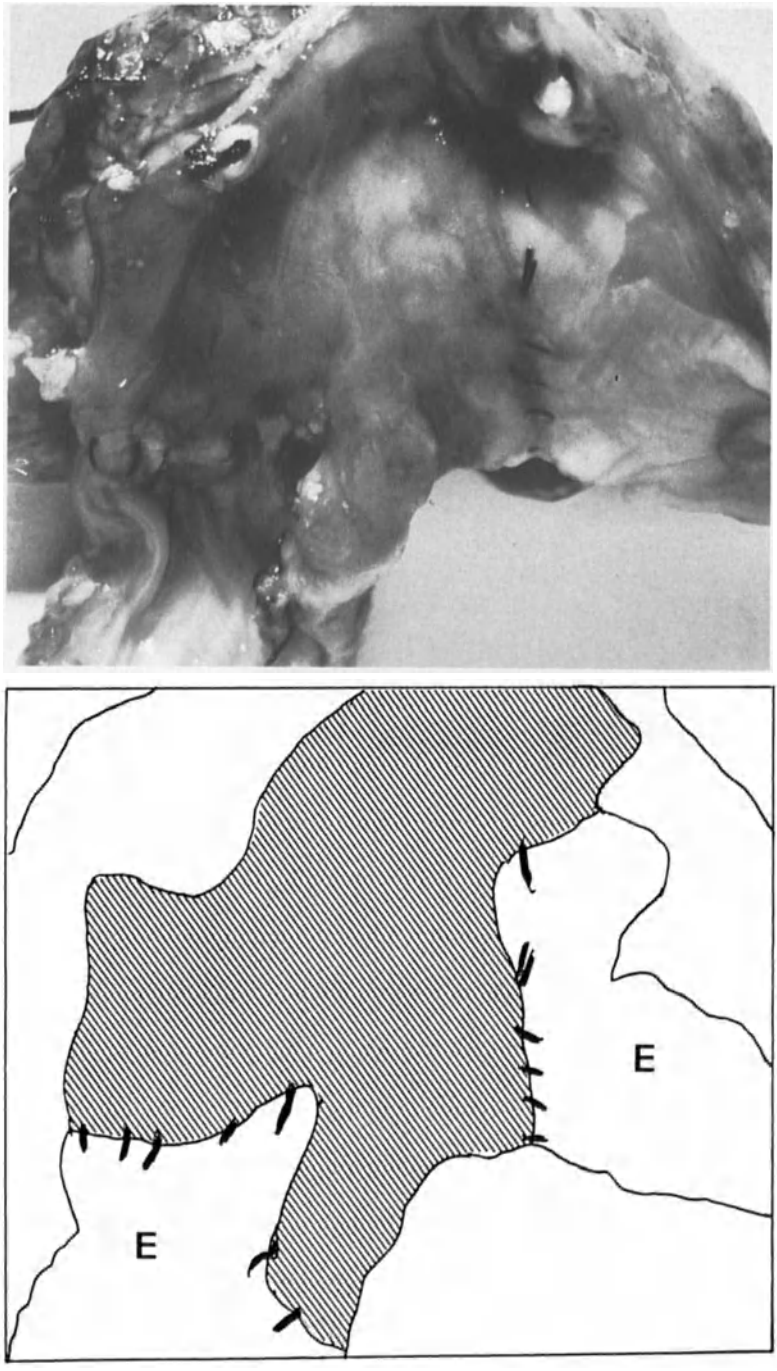

Fig. 5. a Vital jejunal autotransplant at time of killing, 2 weeks postoperatively. **b** Autotransplant represented by shadowed area. *E,* esophagus

was killed the transplant was found to be completely intact, with well-healed anastomoses (Fig. 5). The microvascular anastomosis to the aorta had also healed well.

Another piglet lived for 6 weeks without evident problems but suddenly died of a hematemesis due to a fistula between the aorta and the esophagus. Several piglets lived, apparently well, for between 1 and 3 weeks but then died of transplant necrosis.

Conclusions

The operative method tested has a number of theoretical advantages. It allows the use of all esophageal tissue available and substitution of the defect with a vital and elastic tissue. In addition, the huge loss of bowel associated with colonic interposition procedures is avoided.

The present study has shown that it is possible to correct a thoracic esophageal defect with a jejunal autotransplant using a microvascular anastomosis to the aorta. With the animals and experimental conditions used, the method entailed a high incidence of complications and mortality, but it is not at all sure that this must be the case with human beings who have access to intensive care. However, we do not think the time has yet come to use this kind of procedure in infants; we are continuing to work experimentally in order to find simpler and safer methods for the microvascular end-to-side anastomosis to the aorta.

Summary

An experimental method is reported for bridging defects of the thoracic esophagus, using a free jejunal graft connected to the aorta by microvascular anastomoses. The advantages of this method are that defects are bridged by vital and well-vascularized tissue and that only short bowel segments are required. Drawbacks seen in animal experiments are a relatively difficult operative technique and a high complication rate. This high complication rate might be lowered in infants under intensive-care conditions.

Résumé

Une méthode expérimentale pour ponter les pertes de substance de l'œsophage thoracique est décrite; cette méthode utilise un greffon jéjunal libre dont les vaisseaux sont connectés à l'aorte par anastomoses microchirurgicales. L'avantage de cette méthode réside dans le fait que les pertes de substance sont pontées par un segment intestinal bien vascularisé court. En revanche, il s'agit d'une technique opératoire relativement difficile avec un haut pourcentage de complications qui pourrait être abaissé chez les enfants sous soins intensifs.

Zusammenfassung

Es wird berichtet über die Überbrückung von Defekten im Bereich des thorakalen Ösophagus mit einem freien Jejunumtransplantat, unter Benutzung mikrovaskulärer Anastomosen zur thorakalen Aorta. Der Vorteil dieser Methode ist, daß der Ösophagusdefekt durch lebendes, gut durchblutetes Gewebe überbrückt werden kann und daß nur ein relativ geringer Darmanteil aus dem Abdomen geopfert werden muß.

Als Nachteile dieser Methode erwiesen sich im Tierexperiment eine relativ schwierige operative Technik und eine hohe Komplikationsrate. Unter Intensivpflegebedingungen könnte diese Komplikationsrate beim Menschen jedoch sicher gesenkt werden.

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Management of Prenatally Diagnosed Congenital Malformations – Actual Problems and the Importance of an Interdisciplinary Team Approach

U. G. Stauffer[1]

Modern methods of prenatal diagnosis – especially ultrasonography, amniocentesis, and fetoscopy – provide us with early information concerning the clinical state of the fetus. Reports in the lay press of sensational intrauterine surgery have made the public aware of these possibilities and have led to considerable, sometimes very controversial discussions. Intrauterine therapy has been both hailed as a great advance and condemned in various articles in the lay press as well as in scientific journals. This highlights the fact that there is a considerable amount of uncertainty about the actual value of these new methods and their practical consequences today. Early recognition of congenital malformations in utero may be the basis for many important decisions regarding the future of mother and child. The same prenatal diagnostic methods not only help the future child but also allow the physician to consider selective termination of pregnancy. Termination of pregnancy is certainly justified, for instance, in cases of antenatally diagnosed anencephaly. In the great majority of cases, however, the situation is less clear cut and may lead to serious ethical, moral, religious, and even legal disputes. The complex problems of prenatal diagnosis and their consequences can therefore not be discussed in isolation in an ivory tower of modern medicine. They have to be put within the social, socioeconomic, ethical, and religious conditions of the western society of today.

The Physician and the Current Social Context – When Does the Fetus Become a Patient?

The complex situation is schematically shown in Fig. 1. Modern society behaves paradoxically in many ways indeed. On the one hand we have, for instance, the techniques of artificial extrauterine insemination, techniques which are considered a great advance and which are supported with enormous financial subsidies. On the other hand, there is an ever increasing trend toward abortion. It is estimated that the number of abortions compared with the number of live births is about 1:1 in countries where birth control is either forbidden or not practiced because of religious reasons. In Italy, the relation is supposed to be even 3:1. On the one hand, the wanted child is one of the greatest treasures of life; on the other, the unwanted child is thought by many to have no intrinsic value at all. A profes-

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sor of theology discussing this strikingly paradoxical behavior of our society pointed out that the fetus has been degraded today to a sort of collector's item.

Modern advances in our knowledge of embryology as well as in the techniques of prenatal diagnosis and possible therapy have revitalized the question of whether the unborn fetus is already a human being. Current law holds that a person begins at birth. Future law may state that the individual becomes a person at the moment of conception. This has already been debated in the American congress with the so-called Helms-Hyde bill of 1982. This new concept is, of course, radically different from the concept of many philosophical teachings since antiquity and different from the dogma of many theologians, who as late as in the 19th century taught that the fetus becomes a human being with a soul only during successive phases of pregnancy. A boy was thought to be a human being from the 40th day after conception on, girls from the 80th or 90th day on. This grotesque time limit is as arbitrary as other current ideas, for instance that the fetus becomes an individual at the moment of nidation, or at the moment he becomes indivisable around 14 days after conception, or at the point where brain development starts, or by the time of recognition of measurable brain activity, etc.

In former times these opinions were only of theoretical and philosophical value, but today they have grave practical consequences. The question that arises for the parents, especially the mother, and for the physician is whether the fetus is already a patient with full human and legal rights. Looking at all these problems we have to realize that in many countries, for instance the Federal Republic of Germany, Great Britain, and the United States, every woman can freely decide whether she wants an abortion, even of healthy child, before the 16th, 20th, or

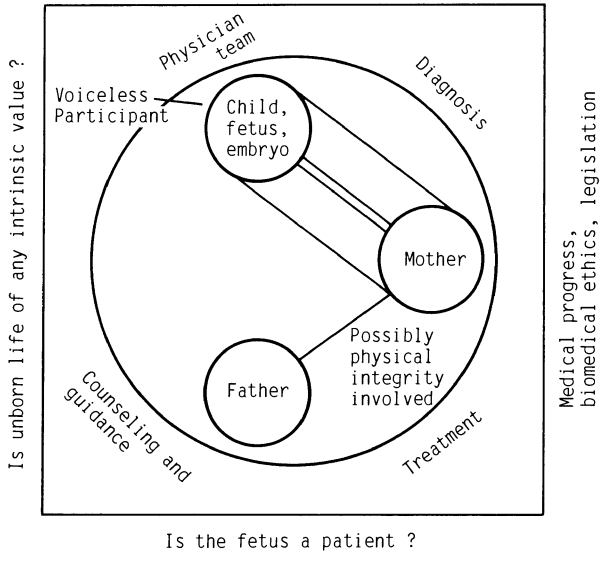

Fig. 1. Possible consequences and problems of prenatal diagnosis in today's social context

24th week of pregnancy respectively. The everyday work of a gynecologist and obstetrician in a large hospital may therefore easily consist of three to four terminations of pregnancy in one morning's operating session. It must therefore be a sort of paradox for an obstetrician when he is suddenly confronted with a mother willing to have her child who has a more or less severe pathological finding on ultrasonic examination in the same period, between the 16th and 24th week of pregnancy. Certainly, the mother is the primary patient for the obstetrician. It is the mother who has to bear the consequences of any decision made in cases of fetal congenital malformations. On the other hand, the unborn child is also the responsibility of the obstetrician, and conflicts may suddenly arise between the mother's rights and interest and the possible rights of the fetus. Initially I raised the question of whether the fetus is already a patient in the true sense with full rights. Regarding our abortion laws and the everyday practice of abortion, I am sure we will have serious doubts. In the complex situation of intrauterine diagnosis of congenital anomalies, rational advice on therapy will therefore require even more extensive interdisciplinary discussions.

Intrauterine Diagnosis of Congenital Malformations of the Fetus: Consequences for the Mother

Table 1 attempts to show the possible consequences for the mother when a congenital malformation of the fetus is discovered in utero. There are a number of advantages but also a number of disadvantages. In the first trimester, exact recog-

Table 1. Possible consequences of intrauterine diagnosis of fetal malformations for the mother

Advantages	
First trimester:	– Recognition of dead fetus
Second trimester:	– Elimination of the possibility of severe malformations
	– Feeling of security
Third trimester:	– Feeling of security if optimal treatment can be carried out for correctable malformations
	– Termination of hopeless pregnancies
	– Better psychological management of the mother

Disadvantages

- Psychological strain for the parents if a malformation has been diagnosed: Diagnosis may be certain, questionable, temporary changes possible
- Having to live with malformed fetus
- Difficulties of deciding on
  - Abortion
  - Induction of premature labor
  - Cesarean section in the interest of the child

- Mother forced to undergo treatment (cesarean section in spite of mother's wish to the contrary!)

nition of a dead fetus avoids unnecessary therapy, and anxiety and false hopes on the part of the mother. In the second trimester, the exclusion of severe malformations, chromosomal aberrations incompatible with life, etc., are of prime importance. The certainty that the child is developing normally could also be an additional psychologically important advantage for the parents. However, there are already some cautious reservations concerning the accuracy of intrauterine diagnosis. For instance, we have seen a child who was born with severe malformations of the limbs, rectal atresia, and complex cardiac malformation who died a few hours after birth. Ultrasound investigation was carried out in a large clinic during the 28th week of pregnancy and had repeatedly shown a so-called normal fetus!

Such drastic mistakes due to incompetence of the ultrasonographer could probably be avoided with the three-step concept proposed by Hansmann in Bonn in 1981. This concept envisages a routine ultrasound investigation, usually in the outpatient office of an obstetrician. If there is any suspicion of pathological changes the mother should then be sent to a competent obstetrical hospital that has experts in ultrasonography. If a definite diagnosis is still not possible, the patient should be sent to a highly specialized diagnostic center as a third step. According to Hansmann, three to six such special centers would be sufficient for the Federal Republic of Germany (Hansmann 1981). It must, however, be stressed that even under ideal diagnostic conditions the layman's expectation that today all important congenital malformations can be diagnosed prenatally is not realistic. We recently witnessed the tragedy of parents who had waited for a child for 10 years and had been regularly assured that the unborn child would be normal by serial, competent ultrasound examinations. At birth, however, the parents were faced with an infant with an open myelomeningocele. At one of the most expert centers of western Europe, in Bonn, Hansmann could make the diagnosis of myelomeningocele in only 17 of 21 cases.

Intrauterine diagnosis in the last 3 months of pregnancy allows for the planning of an optimal prenatal management of children with correctable malformations. On the other hand, hopeless pregnancies can be terminated where survival would be impossible, such as cases of Potter's syndrome.

There are also a number of definite disadvantages (Table 1). Early recognition of an intrauterine malformation produces a severe psychological strain for the mother. Nevertheless, the modern mother, aware of the diagnostic methods available, will request exact information after the first ultrasound examination. It is extremely difficult to decide what and how much should be told to the mother at this early stage. The problem is even more difficult considering the fact that malformations are difficult to diagnose during early pregnancy and that some of them, such as hydronephrosis and hydroureter, may even disappear during the coming weeks and months. Naturally, the strain on the mother becomes even greater when a decision must be made to perform an abortion or premature induction of labor, for instance for a fetus with Potter's syndrome, or whether the mother should undergo a cesarian section.

The disadvantages mentioned in Table 1 can be avoided to a certain extent by competent and understanding discussion between the physician and the patient, but

they cannot be completely eliminated. Whether and when the advising specialists, including the pediatric surgeon, should be asked to discuss the problem with the parents depends on the decisions and customs of the local team.

We strongly believe that it is ethically wrong to force a mother to undergo treatment in the interest of the child against her will. We realize, however, that in extreme cases the interests of mother and child may be diametrically opposed. There have already been two decisions by American courts of justice forcing a pregnant woman to have a cesarean section against her will in order to save the fetus, stating that the fetus' right to live was more important than the risk of an operation to the mother. These decisions have been severely criticized by groups in Chicago and Boston, and the criticism has been published under the title "Forced Cesareans: The Unkindest Cut of All" (Annas 1982). We agree with Pringle (1984) that as long as fetal therapy is available only through the mother, the maternal right to refuse treatment must remain sacrosanct.

Intrauterine-Diagnosed Malformations – Possible Consequences for the Child

The possible consequences for the child are shown in Table 2. Definite advantages are the screening of patients at risk, the treatment of mother and child at a competent center, the choice of the best time and way of birth, and treatment by an interdisciplinary team before, during, and immediately after birth. Intrauterine therapy, especially intrauterine surgery, however, is only beginning, and in spite of many published reports it still has an experimental character. Elias and Annas have remarked: "Nevertheless it seems fair and accurate to conclude that all of the procedures described must currently be considered experimental" (Elias and Annas 1983). Harrison, who is certainly the person with the greatest experience in fetal surgery, wrote: "Treatment of the fetus with a potentially correctable defect is promising but still experimental" (Harrison et al. 1982a). In a discussion at the same meeting he said: "I think we all should maintain a healthy scepticism about fetal treatment. We just don't know enough yet about the efficiency, feasibility or safety of this method of treatment. I am scared to death that unfortunate publicity and unrealistic expectations will lead to ill-advised attempts at treat-

Table 2. Possible consequences of intrauterine diagnosis of a fetal malformation for the child

– Better management during the perinatal period through
– Screening of mother and child at risk
– Management by a team during and after birth
– Choice of best possible time for birth
– Choice of best possible type of birth (vaginal or cesarean section)
– (Intrauterine surgery)
– Abortion

ment. The consequences of irresponsible application will fall not only on the fetus and mother but on the development of a promising field" (Harrison et al. 1982b).

In Table 2, abortion as a possible consequence for the child is mentioned beneath the line. Abortion, and therefore nonexistance, can by definition not be mentioned as being in the interest of the child. It may, however, be considered the right measure in the interest of the parents and/or society, for instance in a case of anencephaly or Potter's syndrome.

The Interdisciplinary Team – the Contribution of the Pediatric Surgeon

Until a few years ago gynecologists tended to ask only the geneticist for advice when deciding whether it was reasonable or necessary to terminate a pregnancy. During the past few years the neonatologist has penetrated into delivery rooms and has become an advisor and helper during and immediately after delivery.

The problems of congenital malformations discovered in utero can only be solved by a much larger team of specialists, however, composed of the obstetrician, the ultrasonographer, the neonatologist, the pediatric surgeon, and, if necessary, neurosurgeons, cardiac surgeons, etc. In special cases it may also be advisable to include embryologists, physiologists, and even an independent ethical committee. Such an ad hoc workshop group including the pediatric surgeon has existed in Zürich for the past 4 years. The help to be given by pediatric surgeons to such an interdisciplinary team can be seen from Table 3. They inform their colleagues objectively about the possible corrective methods, and the immediate and long-term results in such patients. They must also advise their colleagues whether a malformation can be corrected only incompletely or not at all and of the consequences in later life, degree of handicaps, etc. They can base their advice on their own experience or on the published literature. The pediatric surgeon is therefore not only a responsible advisor about objective data for the obstetrician, but also acts perforce as lawyer for the unborn child. The pediatric surgeon should also help in ascertaining the possible gain brought by early intrauterine diagnosis and its effect on therapy. Our experience is as yet too small for us to be dogmatic about these problems, and only first impressions can be related here. A few examples may help to illustrate these problems.

Table 3. Contributions of paediatric surgeons

1. Information about modern methods for correcting malformations (in cases of *postnatal* diagnosis!)
   - Early results
   - Late results (after years!!)
2. Information about possibly not or only partially correctable malformations and their consequences for later life
3. Evaluation of a possible advantage of *prenatal* diagnosis for the child in case of
   - immediate postnatal treatment (fetal surgery)

The diagnosis of *gastroschisis* can be made prenatally with considerable certainty, but the obstetrician often has no clear idea about modern therapeutic methods for treating this malformation. The changes in the prognosis for gastroschisis during the past few years can be seen in Table 4. This shows how important it is for the obstetrician to be informed about the most recent scientific literature. Before 1968 only two of ten children with gastroschisis survived in Zürich; between 1970 and 1981 13 of 14 survived. Similar results were obtained in Los Angeles, Chicago, and Columbus. Considering these good pediatric surgery results, abortion for this condition should therefore be out of the question. A mother with a child with gastroschisis must, however, be admitted to a correspondingly competent clinic. Early induction of labor is not indicated, but today cesarean section is usually advised, as contamination of the prolapsed intestine can thus be avoided.

Table 4. Chances of survival for gastroschisis patients

Zürich, 1952–1968	20% (2/10)
Zürich, 1969–1980	90% (11/12!!)
Columbus, 1973	70%
Chicago, 1974	84%
Los Angeles, 1976	89%

Table 5. Omphalocele – chances of survival of 128 patients[a]

Groups at risk	Patients *(n)*	Chances of survival
Group A	65	88%
Group B	48	48%
Group C	15	13%
Total	128	58.5%

[a] Including 12 with trisomy 13/15, 17/18, 21

In children with *omphalocele* the situation is completely different. As is generally known, these children suffer frequently from additional malformations, mainly of the GI tract and the heart, and omphalocele is often associated with prematurity and underweight. The example of omphalocele shows how important it is for the obstetrician to realize that malformations which grossly resemble each other can have a very different prognosis. Pediatric surgeons have therefore long classified such patients into risk groups according to the presence or absence of concomitant severe malformations, prematurity, and underweight. Our obstetric colleagues are usually not familiar enough with these risk groups and modern postnatal pediatric surgical management.

Table 5 shows survival according to such risk groups in 128 children with omphalocele in Liverpool. Statistically, a child in group A had an 88% chance to survive, a child in group C a chance of only 13%. When discussing such statistics in our interdisciplinary team, it is the duty of the pediatric surgeon to point out that there are cases in group C in which the pediatric surgeon – together with the neonatologist and the geneticist – did not advise attempting surgical correction, for instance, for patients with chromosomal aberrations and/or other uncorrectable malformations.

I would like to illustrate the importance of the presence of a pediatric surgeon on the interdisciplinary team by means of the tragic example of a fetus with

omphalocele. Only 6 years ago, a fetus with an isolated small omphalocele, without additional malformation and without chromosomal aberration, was aborted at a large Swiss University obstetric clinic; the decision was based on statistics of late results in an article published in 1969. The pediatric surgeon was not included in this decision and the advice was given by the geneticist alone.

Possibilities of Intrauterine Therapy at Present

As antenatal diagnosis of congenital malformations during the second trimester has become possible, the obstetric team has been more inclined to ask the pediatric surgeon whether intrauterine therapy could improve the chances of the fetus. Here a critical appraisal of the situation by the pediatric surgeon is very important. Altogether there are 92 conditions that have so far been diagnosed antenatally; 17 of them are surgical cases but only a few are possible candidates for intrauterine surgery. The most important patient groups are children with diaphragmatic hernia, those with severe obstructive uropathies, and those with hydrocephalus. Harrison et al. (1981) and Pringle (1984) have shown that diaphragmatic hernia can be produced experimentally in fetal lambs and can later on be operated successfully in utero, but up to the present time no intervention has been attempted in the human fetus. Harrison himself pointed out that there are still too many unknown factors involved, and that the time for application of these techniques in human beings has not yet come. Considering the high risk of fetal surgery, it must be realized that there are no selection criteria today for determining which fetuses with diaphragmatic hernia would really be unable to survive without intrauterine operation and which could survive when operated on in the usual way just after birth. However, biological studies in this field have already provided us with much important information. For instance, it is known that the bronchial tree is already completely formed by the 16th week of intrauterine life, and that during the 23rd week the alveoli begin to develop, whereas the capillaries are not yet present.

The situation of fetuses with hydrocephalus diagnosed in utero is similar. There are as yet no definite data on how to select patients for intrauterine drainage for hydrocephalus. There are also no generally accepted criteria for evaluation of a possible successive therapy. It is important that the pediatric surgeon points out that the width of the cortex cerebri is not representative of later mental and intellectual development of the child. Nevertheless, Clewell and collaborators of Denver, Colorado drained the lateral ventricles of a fetus with marked hydrocephalus into the amniotic cavity in the 24th week of pregnancy (Clewell et al. 1982). The drainage worked until the 32nd week. The authors believe that they might possibly improve the prognosis for such patients, and they have operated on two further fetuses with hydrocephalus since June 1982. They are well aware of the controversal nature of their experimental work, however. They therefore approached the Human Subjects Committee of the University of Colorado before operating, asking them to appoint an independent advocate for these fetuses.

Theologists and neonatologists were appointed as independent advisors and advocates. In discussing the intrauterine therapy of hydrocephalus it is up to the pediatric surgeon to point out all these controversies, uncertainties, and ethical reservations, and to stress the still experimental character of such operations.

Among the prenatally diagnosed malformations, fetuses with obstructive uropathies form by far the largest group. More than half of all articles about prenatal diagnosis deal with this subject. In about 90% of cases the fetal kidneys can be demonstrated exactly by ultrasonography as early as the 17th–20th week. The great advances in ultrasonographs of the second and third generation have made it possible to visualize some anomalies of the urinary tract in the 12th–15th week of pregnancy. The human fetal kidney starts to produce urine as early as the 11th week. Early obstruction during the first month of pregnancy leads to renal dysplasia, obstruction during the second half of pregnancy to various degrees of hydronephrosis and hydroureter. One of the most severe obstructive uropathies can be found in children with urethral valves. Of our 12 patients with congenital urethral valves treated immediately after birth or several months after, one has died; six of the 11 surviving patients suffered from renal insufficiency during the first few years of life. These very depressing results have been confirmed in a collective review of the British Association of Paediatric Surgeons in the United Kingdom. Under these circumstances, it is only logical that the question has been raised as to whether early drainage of the bladder into the amniotic cavity would not improve renal function. The same question is raised in connection with other severe obstructive uropathies which may finally lead to Potter's syndrome. Extensive pathophysiological studies of fetal lambs and monkeys with obstructive uropathies have been carried out by Harrison and his collaborators in San Francisco. Based on these animal experiments, drainage of the bladder into the amniotic cavity was performed by this group in 26 human fetuses up to 1983 (Harrison et al. 1982c). The results results are on the whole disappointing. Even in a fetus in which bilateral cutaneous ureterostomies were carried out as early as the 21st week of pregnancy, advanced renal dysplasia was found at autopsy (the child died on the first day of life of an unrelated cause). Bellinger (1982) described a fetus with urethral valves that died during the 18th week of pregnancy and already had severe renal dysplasia. Modern experimental and clinical results therefore seem to show that the really severe types of renal dysplasia occur so early during pregnancy that all intrauterine surgery practiced today comes too late. When discussing fetal surgery in obstructive uropathies with obstetricians and neonatologists, it must be pointed out further that it is not possible today to measure the function of the fetal kidney with accuracy. It is therefore also so far impossible to objectively evaluate results obtained by intrauterine surgery in such cases (Kramer 1983).

The great advantage of prenatal diagnosis of genitourinary malformations today therefore lies mainly in the possibility that patients at risk can be recognized early on, and severe malformations can be corrected by surgery in the immediate postnatal period, perhaps – in some selected cases – following a planned cesarean section after the 32nd week of pregnancy. These early interventions prevent further harm to the kidney from additional severe urinary infections.

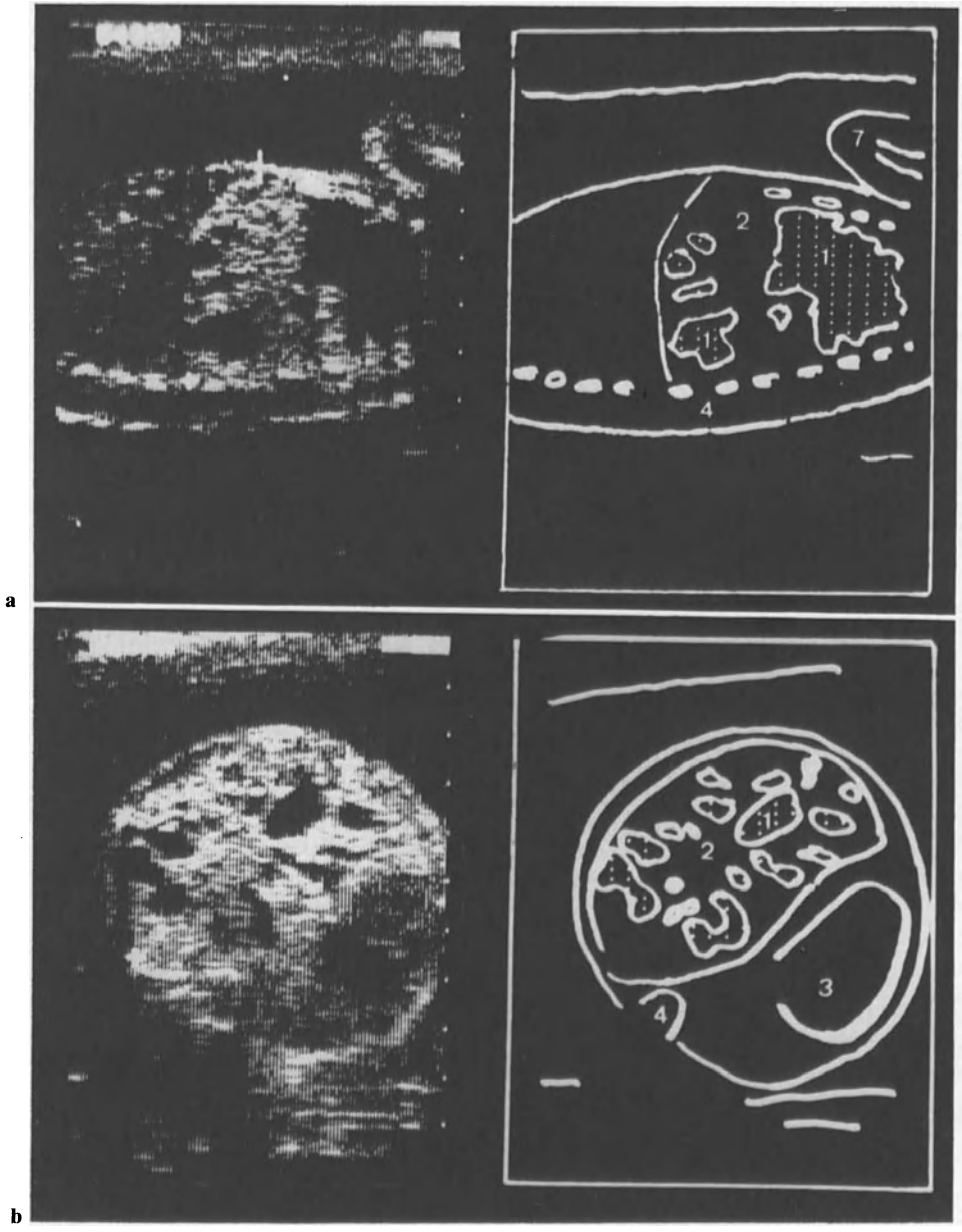

Fig. 2a, b. Ultrasound findings at 27 weeks of gestation: **a** longitudinal section; **b** cross-section. *1,* Normal lung tissue; *2,* cystic adenomatoid lung; *3,* displaced heart; *4,* vertebra/spine; *7,* arm

The modest results of fetal surgery may inspire doubts about whether this complex and expensive research is really valuable and promising for the future. Considering all the new basic knowledge in embryology and pathophysiology, and the understanding of congenital malformations gained by researchers in this field, I feel that these doubts are not justified. I think, however, that research concerning fetal surgery should be restricted to a very small number of centers. A good example is the Fetal Treatment Program headed by Harrison, who has put together a large team of basic researchers, working jointly with clinicians, obstetricians, pediatric surgeons, ultrasonographers, neurosurgeons, anesthesiologists, and ethicists. The present state of fetal surgery has recently been critically reviewed by this group (Harrison et al. 1984).

According to Pringle (1984), some of the most urgent prerequisites for fetal surgery – besides developing and exploring the necessary experimental techniques of creating defects in animal models in utero and then correcting them before the fetus is born – are as follows:

1. The gathering of prospective data on the incidence and prognosis of various anomalies
2. The development of reliable means of suppressing premature labor before any major fetal surgery with unduly high risk can be undertaken
3. The development of an artificial placenta, "the conductor of the entire endocrine orchestra of pregnancy" (Beaconsfield et al. 1980)

The problems of placental transfer have been outlined by Young (1979) and Morriss (1981). We have to realize, however, that any fetal intervention can finally be carried out only through the mother. Even if an artificial placenta could be developed – and many researchers are working in that direction – one would still need to withdraw the fetus from the mother by a cesarian section before connecting the fetus to the artificial placenta!

New developments in all fields of surgery have always been criticized and regarded skeptically. In 1881, Billroth openly stated that a surgeon who tried to suture a heart wound should lose the respect of all his colleagues. Nevertheless, modern heart surgery has developed brilliantly since then! We all hope that fetal surgery may develop in a similar way! However, it is the duty of the pediatric surgeon to point out that really uncontestable success with fetal surgery has, with a few exceptions, not yet been achieved, and that it will probably take years of basic and clinical research before a safe, broad clinical application is possible.

On the other hand, I would like to illustrate the current benefit of prenatal diagnosis and an interdisciplinary team approach for perinatal and postnatal care by a case of adenomatoid malformation of the lung that I personally handled. Ultrasound examination of a 20-year-old woman during the 27th week of pregnancy showed a multicystic, partly solid tumor in the left hemithorax of the fetus (Fig. 2). The ultrasonographer, the pediatric radiologist, and the obstetrician discussed the findings together and suspected an adenomatoid cystic malformation of the left lung. No additional malformations were discovered and the fetus had developed normally so far. The pediatric surgeon on the interdisciplinary

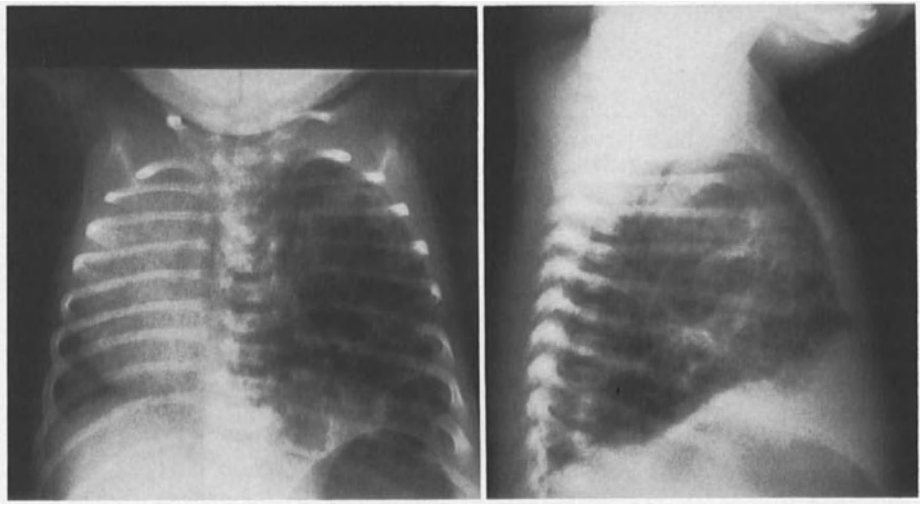

Fig. 3. X-ray taken immediately after birth. Multicystic lesion filling the left hemithorax, with massive displacement of the heart and the mediastinum to the right and compression of the right lung. Abdomen filled with gas-containing loops

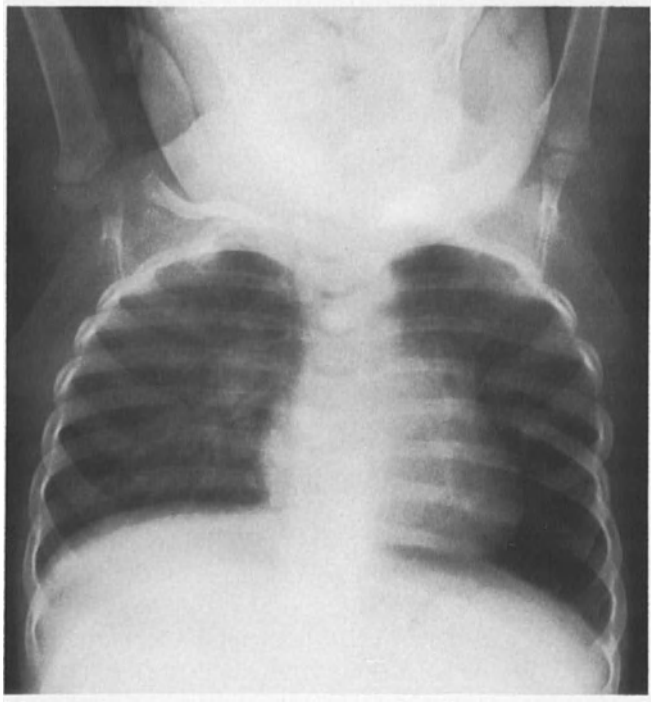

Fig. 4. X-ray taken on the 10th postoperative day

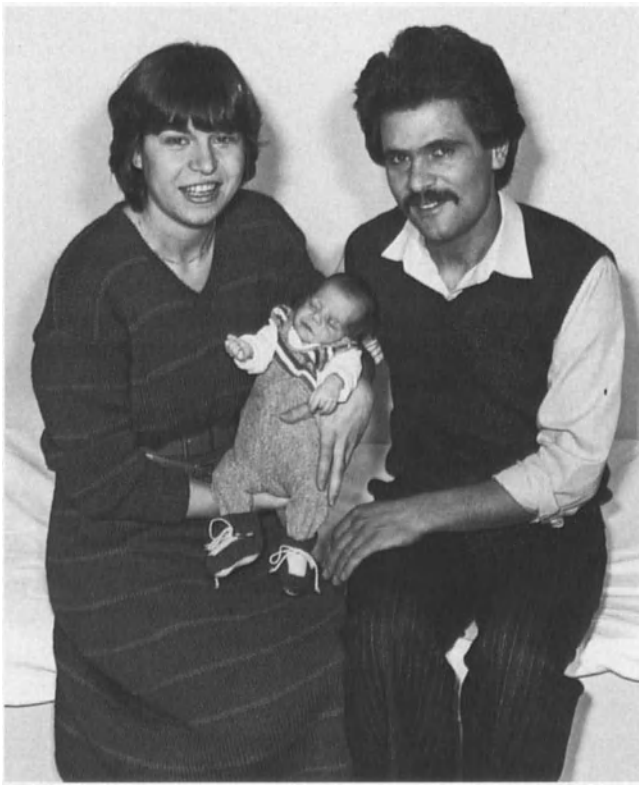

Fig. 5. The little patient and his family

team was consulted about possible management and prognosis. He pointed out that this rare malformation usually affects only one lobe of the lung; in only two of 31 cases reported in the literature were two lobes affected. It was thought that prompt surgical intervention just after birth should be successful, and it was decided to allow the pregnancy to go to term. The mother was told about the suspected diagnosis by the team. Further ultrasonographic examinations were carried out at regular intervals. They showed that there was no excessive growth of the malformation in comparison with the general growth of the fetus. A cesarean section at term was considered. Adequate postnatal management would, of course, have been facilitated by a planned cesarean section. However, 30% of the alveolar fluid is normally expressed when the thorax passes through the birth canal. This was regarded as an important natural contribution to the postnatal adaption, especially for this child. We therefore decided on a normal vaginal birth at term. As a consequence, a pediatric surgical team was constantly on call during the last 3 weeks of pregnancy for a possible emergency operation. When labor began, an operating room was reserved in the Children's Hospital. A pediatric anesthetist and a neonatologist were present at birth. Figure 3 is the radiograph of the thorax immediately after birth with the characteristic adenomatoid mal-

formation of the left lung and massive shift of the whole mediastinum towards the right. The malformed left lower pulmonary lobe was resected. Figure 4 shows the radiograph 10 days after operation, Fig. 5 the happy parents. Today the child is 4 years old and has developed completely normally. This is the first patient with a prenatally diagnosed pulmonary adenomatosis, successful operation, and long-term survival.

Summary

Today's methods of prenatal diagnosis, i.e., ultrasound, amniocentesis, and fetoscopy, allow for early recognition of abnormalities in the fetus. Trials of surgery on human fetuses are widely discussed in scientific as well as lay journals; they are accepted with enthusiasm by some and severely critisized as unethical by others.

This report deals with the modern concepts of prenatal diagnosis and possible therapy in the light of current general social context. Some of the arising controversies and ethical problems are shown. The consequences of prenatal diagnosis of congenital malformations of the fetus are separately discussed with reference to the mother, the family, and society on one side and to the fetus itself on the other. The practical question of whether the fetus is already a person or not is seen against its historical, religious, and philosophical background. The necessity for an interdisciplinary team approach in dealing with mothers bearing malformed children – i.e., cooperation of obstetricians, pediatric surgeons, neonatologists, geneticians, neurologists, etc. – is stressed, and the contribution of the pediatric surgeon within this team is discussed in detail, with practical examples given. The present status of intrauterine therapy is summarized and critically evaluated. Finally, an example is given of the ideal team approach in a case of prenatally diagnosed congenital cystic adenomatoid malformation with a successful outcome and long-term survival.

Résumé

Les méthodes actuelles de diagnostic prénatal, en particulier les ultra-sons, l'amniocentèse et la fœtoscopie, permettent le diagnostic précoce d'anomalies chez le fœtus. Des observations où une chirurgie fœtale a été pratiquée chez l'homme ont été relatées dans des revues scientifiques et non scientifiques, acceptées avec enthousiasme par quelques-uns et sévèrement critiquées comme contraires à l'éthique par d'autres.

Dans la communication présentée, les auteurs examinent les concepts modernes du diagnostic prénatal et les thérapeutiques possibles à la lumière du contexte social général actuel. Nombreuses controverses et problèmes éthiques sont démontrées. Les conséquences du diagnostic prénatal de malformations congénitales du fœtus sont discutées séparément, pour ce qui concerne la mère, la famille et la société d'un côté, et le fœtus lui-même de l'autre côté. La question pratique actuelle est

de savoir si le fœtus est réellement une personne ou pas en fonction de considérations historiques, religieuses et philosophiques. La nécessité d'un groupe interdisciplinaire susceptible de prendre en charge la mère portant un enfant malformé apparaît évidente. Cette équipe nécessite la coopération des obstétriciens, des chirurgiens pédiatriques, des néonatologistes, des généticiens, des neurologistes, etc... Dans cette équipe, le chirurgien pédiatrique doit discuter dans tous ses détails l'état pathologique et donner des exemples pratiques de démonstration. L'état actuel de la thérapeutique intra-utérine est résumé et évalué de façon critique. Finalement, les auteurs donnent un exemple d'approche de groupe idéal chez un enfant porteur d'une malformation kystique adénomatoïde congénitale diagnostiquée avant la naissance, avec évolution satisfaisante et survie à long terme.

Zusammenfassung

Die heutigen Methoden der pränatalen Diagnostik, insbesondere Ultraschall, Amniozentese und Fetoskopie, erlauben die frühe Erkennung von angeborenen Fehlbildungen beim Fetus. Berichte über sensationelle intrauterine chirurgische Eingriffe in wissenschaftlichen Zeitschriften und in der Boulevardpresse haben auch zu Diskussionen in der breiten Öffentlichkeit geführt. Von höchstem Lob bis zu schwersten Anschuldigungen gegen verantwortungsloses Experimentieren um jeden Preis reichen die Kommentare zum Thema intrauterine Therapie.

In der vorliegenden Arbeit wird versucht, die heutigen Möglichkeiten der pränatalen Diagnostik und entsprechende Theorien nicht isoliert im Elfenbeinturm moderner Spitzenmedizin, sondern im ganzen sozialen Kontext zu sehen. Die Konsequenzen der Früherkennung von pränatalen Fehlbildungen werden separat für die Mutter, die Familie und die Gesellschaft auf der einen Seite und dem betroffenen Fetus auf der anderen Seite aufgezeigt. Die Frage, ob der menschliche Fetus bereits eine Person mit vollen Rechten darstellt oder nicht, wird aufgrund historischer, religiöser und philosophischer Gegebenheiten diskutiert. Eine klare Antwort steht bislang noch aus. Die Notwendigkeit einer interdisziplinären Zusammenarbeit beim Therapieplan hinsichtlich pränatal festgestellter Fehlbildungen wird unterstrichen. Eine enge Zusammenarbeit zwischen Geburtshelfern, Kinderchirurgen, Neonatologen, Genetikern, Neurologen etc. ist unumgänglich. Der Beitrag der Kinderchirurgen in diesem interdisziplinären Team wird an einzelnen Beispielen erläutert. Die heutigen Möglichkeiten der intrauterinen Therapie werden zusammengefaßt und kritisch diskutiert. Abschließend wird am Beispiel eines Kindes mit einer pränatal diagnostizierten kongenitalen zystisch-adenomatoiden Malformation der Lungen aufgezeigt, wie das praktische Vorgehen am Krankenbett am günstigsten geplant werden kann.

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Prenatal Diagnosis of Fetal Malformations by Ultrasonography

A. KRATOCHWIL[1]

Every obstetrician is aware that most pregnant women are afraid to carry a malformed baby. This fear increases with the approach of the expected date and becomes unbearable in those women who have already given birth to a malformed baby. Formerly, a woman was at the mercy of fate until delivery. Today, with the possibilities of ultrasonography, the situation has changed. The increased scientific interest is reflected by the numbers of published papers dealing with diagnostic procedures to detect such anomalies:

1977	3
1978	34
1979	97
1980	75
1981	155

As we know that only 2%–3% of all deliveries are affected by malformations, and that only 10% of the registered anomalies are due to viral infections and 30% to heredity, whereas more than 60% are spontaneous, a screening program for all pregnancies should be established to detect as many fetal anomalies as possible prenatally. Such a screening program already exists in the Federal Republic of Germany, where all pregnant women are examined ultrasonographically at least twice. The first examination is performed between the 15th and 21st weeks of pregnancy, the second between the 32nd and 36th weeks.

To cope with this huge number of examinations it is necessary to include clinics and hospitals, as well obstetricians in their offices. The screening program is layed out in such a manner that gross fetal anomalies are diagnosed by obstetricians. Patients with certain or suspected anomalies are sent for further evaluation to specialized clinics. Pregnant women with a history of fetal malformation in a previous pregnancy are also sent to such units. Such a concentration is absolutely necessary due to the rarity of these anomalies (see Table 1).

The best time to start with the screening program is between the 15th and 21st weeks of gestation. At every examination ultrasonic biometry is performed. The biparietal diameter is measured to estimate the time of gestation. At the same time the abdominal diameter of the fetus is measured to detect a possible disproportion between the head and the trunk. The entire fetal anatomy is checked as far as is possible with ultrasound, and the amount of the amniotic fluid is

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Table 1. Frequency of some fetal anomalies

Down's syndrome	1 : 600
Cleft palate	1 : 600
Neural tube defect	1 : 1000
Cystic kidney	1 : 1000
Anencephaly	1 : 1200
Hydrocephaly	1 : 1500
Duodenal atresia	1 : 5000
Patau's syndrome	1 : 5000
Hygroma colli	1 : 6000
Umbilical hernia	1 : 10000
Esophageal atresia	1 : 15000
Holoprosencephaly	1 : 16000
Osteogenesis imperfecta	1 : 25000
Prune-belly syndrome	1 : 35000
Achondroplasia	1 : 40000
Klippel-Feil syndrome	1 : 50000

roughly estimated. Signs which must alarm every ultrasonographer to look carefully for malformations are:

1. An- or oligohydramnios
2. Polyhydramnios
3. Disproportion between head and trunk measurements
4. Intrauterine growth retardation
5. Incoordination of fetal movements

If anomalies are detected amniocentesis will be considered, as many malformations are linked to genetic disorders.

Neural Tube Defects

Anencephaly is a malformation which can be quite easily diagnosed, even by not very experienced examiners (Fig. 1). The ultrasonic appearance is so characteristic that it cannot be missed or misinterpreted. In longitudinal scans the trunk ends in an amorphic mass, clearly discernible in the surrounding polyhydramnios. Fetal movements are rare and sluggish, owing to the accompanying neurological lesions (Campbell 1981; Kratochwil and Schaller 1971b; Kurjak 1980; Robinson 1978). The diagnosis can be made by an experienced examiner as early as during the 11th week of gestation (Campbell 1981; Christie 1978). As this severe malformation is incompatible with postnatal life, the only consequence will be termination of pregnancy.

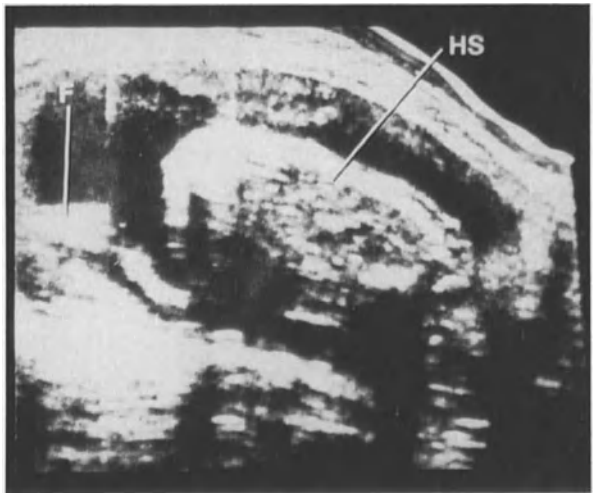

Fig. 1. Anencephaly: the fetal trunk ends in an amorphic mass. Longitudinal scan. *HS*, brain cleft

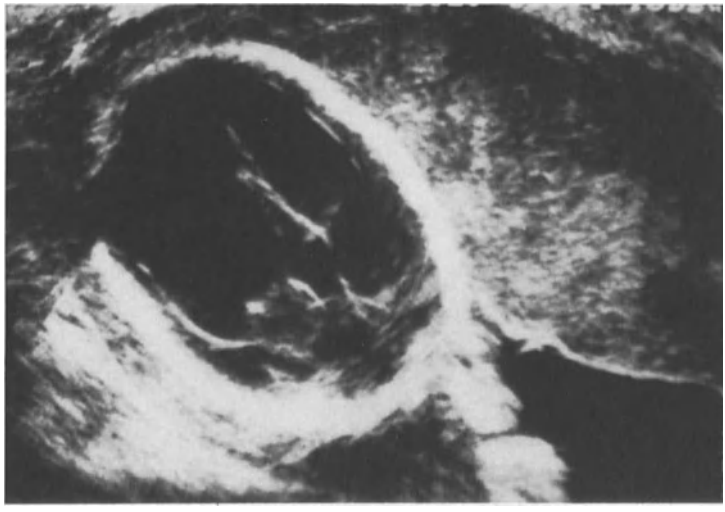

Fig. 2. Hydrocephaly: the picture demonstrates the grossly enlarged ventricles; nearly no brain mantle can be identified. Transverse scan

Hydrocephaly can be detected rather early in pregnancy. Not in all cases the measurement of the biparietal diameter will be far beyond the second standard deviation for the time of gestation. Today the width of the ventricles can be measured and the brain mantle index can be calculated (Campbell 1981) (Fig. 2). The dilated ventricles may increase the biparietal diameter (Fig. 3).

It was recently reported that hydrocephaly might be successfully treated by an ultrasonically guided puncture technique. In this case the ventricles are located by ultrasonic examination and a self-retaining catheter is inserted percutaneously, to

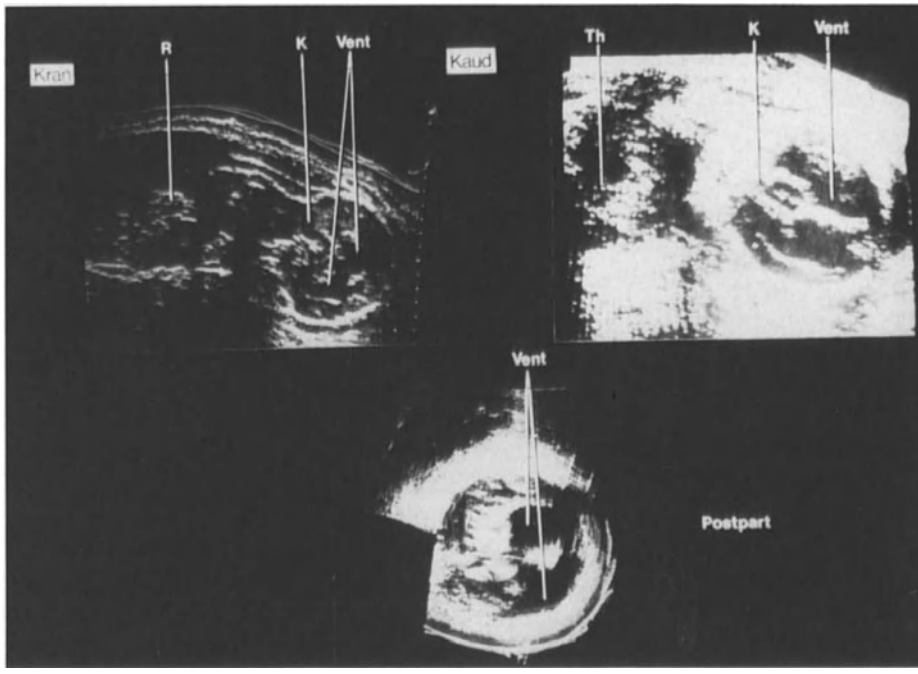

Fig. 3. Hydrocephaly in a small-for-date fetus. Although the ventricles *(Vent)* are enlarged, the biparietal diameter is in the normal range for gestational age. *R,* trunk; *K,* head; *Th,* thorax

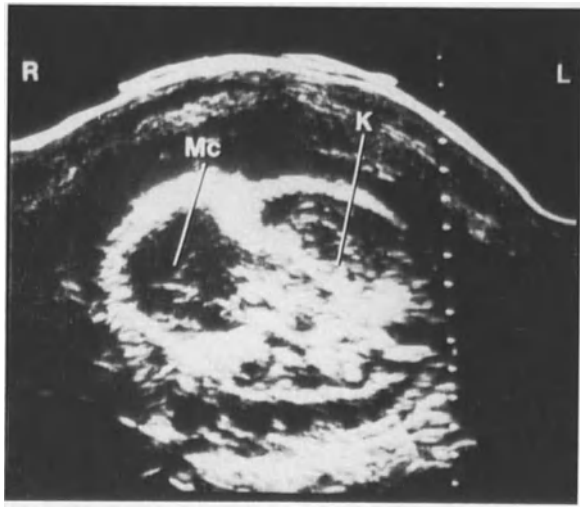

Fig. 4. Myelomeningocele in the neck of the fetus. Transverse scan. *Mc,* myelomeningocele; *K,* head

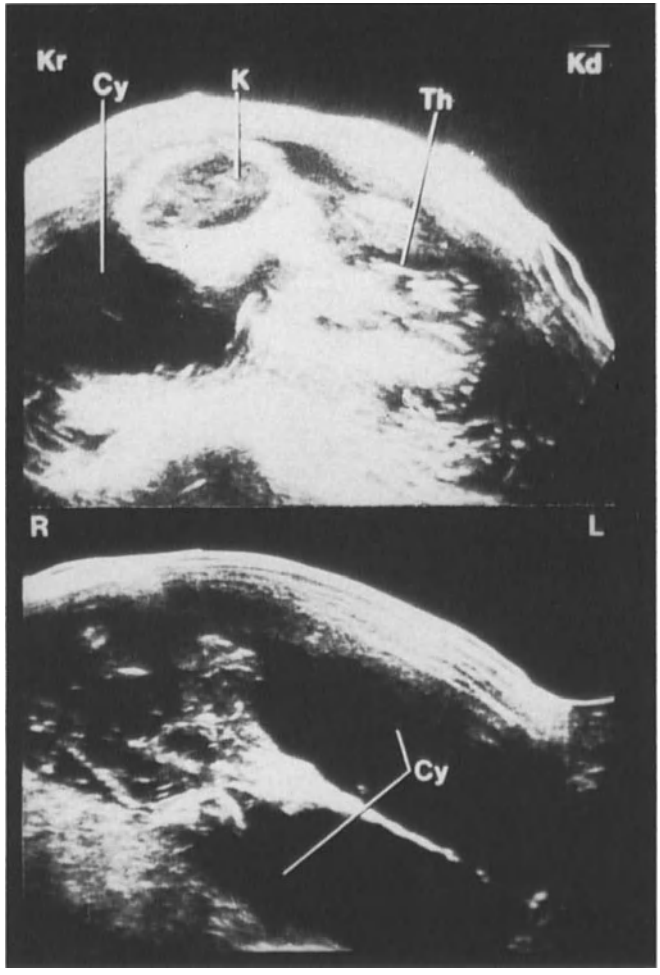

Fig. 5. Hygroma colli. *Above:* fetus in breech presentation. The cystic area cranial to the fetal head may be interpreted as amniotic fluid. *Below:* cystic area is clearly separated by a septum inserting at the fetal neck. *K,* fetal head; *Th,* thorax; *Cy,* cystic mass

drain the ventricle into the amniotic sac (Birnholz and Firgoletto 1981). Such a percutaneous puncture of the ventricles has also been performed in breech presentation to make vaginal delivery possible without any risk for the mother (Borno et al. 1978; Kratochwil and Schaller 1971b).

To detect such lesions as *spina bifida and myelomeningocele* a careful examination is necessary. In some cases where only the vertebral arches are missing a U-shaped deformity can be recognized. The defect has to be localized both in longitudinal and transverse sections (Campbell 1981; Robinson 1978). In case of a larger defect with bulging meninges, a cystic lesion can be outlined in front of the spine. High-frequency transducers also allow the demonstration of neural

tissue in such lesions (Fig. 4). Ultrasonic examination is even superior to X-ray and AFP estimation, as even defects covered with skin can be successfully detected (Christie 1978).

Hygroma colli is seen as a cystic lesion, sometimes divided by septa (Fig. 5). It may be impossible to differentiate this lesion from myelomeningocele. More ventrally located lesions may be of either branchiogenic or thyroid origin (Dunne and Johnson 1979).

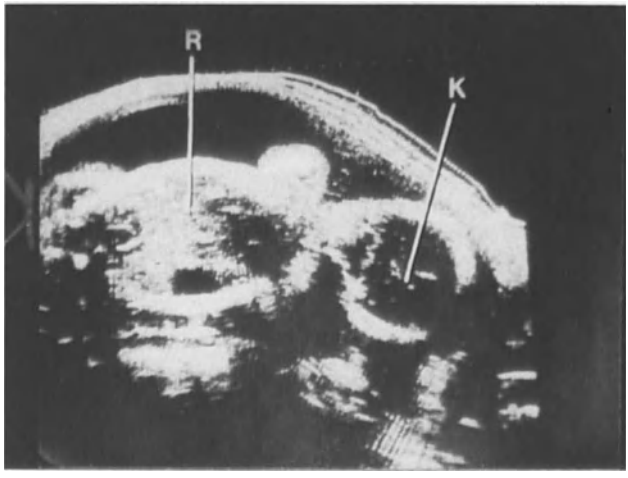

Fig. 6. Cystic adenomatoid malformation. Fetus in cephalic position; a cystic area in the thorax can be clearly identified. *R,* trunk; *K,* head

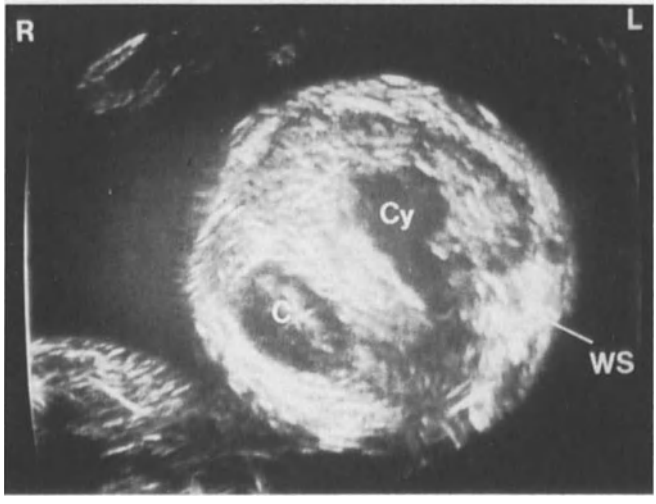

Fig. 7. Transverse scan of the same case as shown in Fig. 6. The cystic mass, shifting the fetal heart *(C),* can be clearly identified. *Ws,* fetal spine; *Cy,* cystic mass

The detection of malformations of the *thoracic organs* is still a problem. Recently an effort was made to detect malformations of the fetal heart. The first reports are very promising, but close cooperation with a pediatric cardiologist is necessary.

Cystic adenomatoid malformation (CAM) of the lung has been diagnosed several times by ultrasonography. This lesion may be of either cystic, solid, or mixed appearance (Figs. 6 and 7). A similar cystic structure may be caused by the fluid-filled fetal stomach in cases of diaphragmatic herniation. Normally, the fetal diaphragm can be identified as a small, linear, anechoic area between the fetal thorax and the abdomen (Fig. 8).

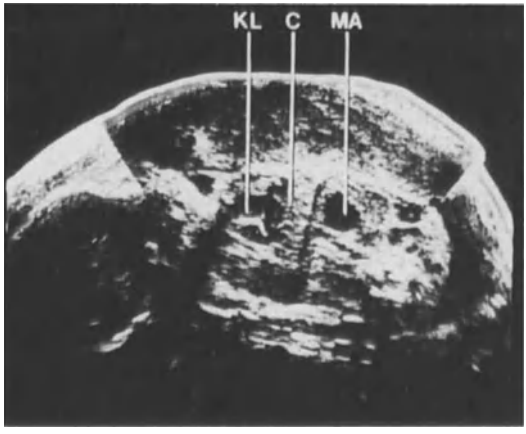

Fig. 8. Identification of the fetal diaphragm. Fetus in breech presentation. Between the thorax, containing the fetal heart *(C)* with valves *(KL)* and the fetal stomach *(MA)* in the abdomen, the anechoic band-like structure of the diaphragm can be seen

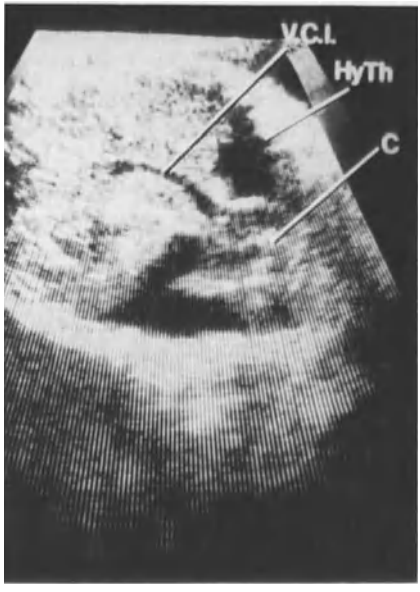

Fig. 9. Pleural effusion. Between the thoracic wall, mediastinum, and diaphragm the pleural effusion is seen as an anechoic area. *C,* heart; *Hy.Th,* hydrothorax; *V.C.I.,* vena cava inferior

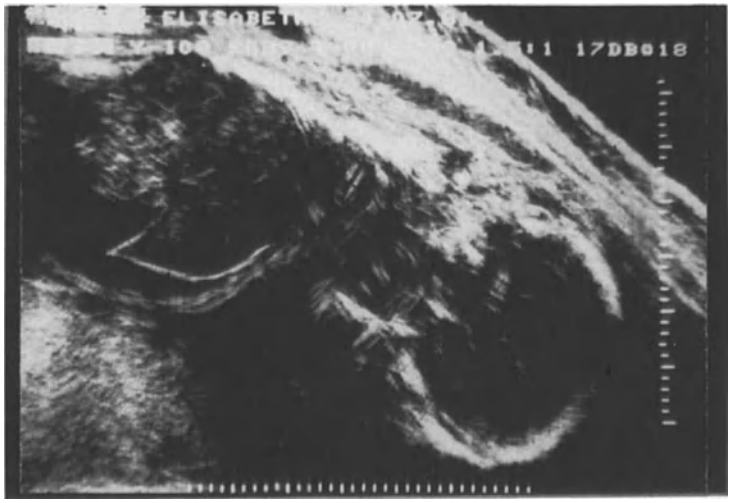

Fig. 10. Ascites: fetus in cephalic position. The sharp-edged fetal liver is separated from the abdominal wall by ascites. Longitudinal scan

Both CAM and a dislocated fetal stomach shift the fetal heart and compress the large vessels, thereby inducing pleural and abdominal effusion (Fig. 7). *Pleural effusion* can be seen as an anechoic space between the thoracic wall, mediastinum, and diaphragm. In the fluid the hypoplastic lungs may be seen to float (Fig. 9). A similar picture is found in ascites, where the liver is separated by the fluid from the abdominal wall (Fig. 10). Isolated effusions in the different compartments of the body can be found as well in cases of cardiac malformation, and even without any detectable reason. The effusions can be successfully treated antenatally by percutaneous, ultrasonically guided needle aspiration.

Detectable Malformations of the Abdominal Organs

Esophageal atresia may be detected by the fact that on repeated examinations the normally fluid-filled, and therefore cystic-appearing stomach cannot be recognized.

In contrast, *duodenal atresia* demonstrates at least two separate fluid-filled areas, depending on the level of the obstruction. Because of these two cystic structures it is called a "double-bubble sign" (Gee and Abdulla 1978; Nikapota and Loman 1979).

Omphalocele can be identified as a tumor in front of the fetal abdomen. Because of the herniation, there is considerable disproportion between the head and the abdomen (Lomas et al. 1979; Niesen and Hansmann 1979). The content of the omphalocele can be either intestinal loops or liver. Both the description of the content and the measurement of the lesion may be extremely helpful in discussion

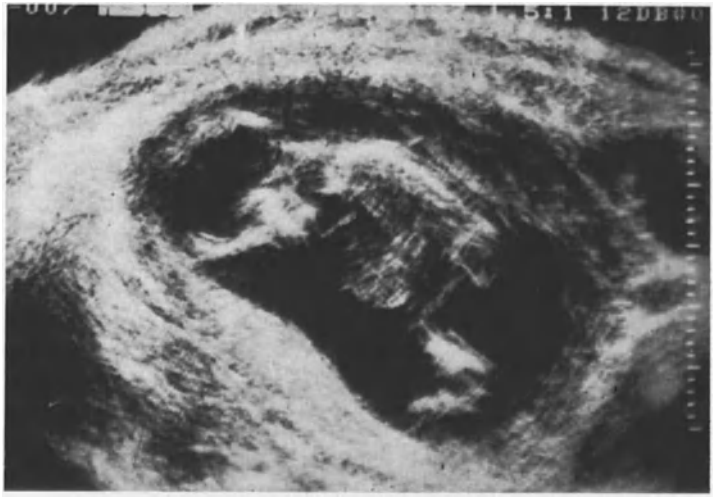

Fig. 11. Omphalocele: fetus in breech presentation, 16th week of gestation. In front of the fetal abdomen the omphalocele is clearly demonstrated

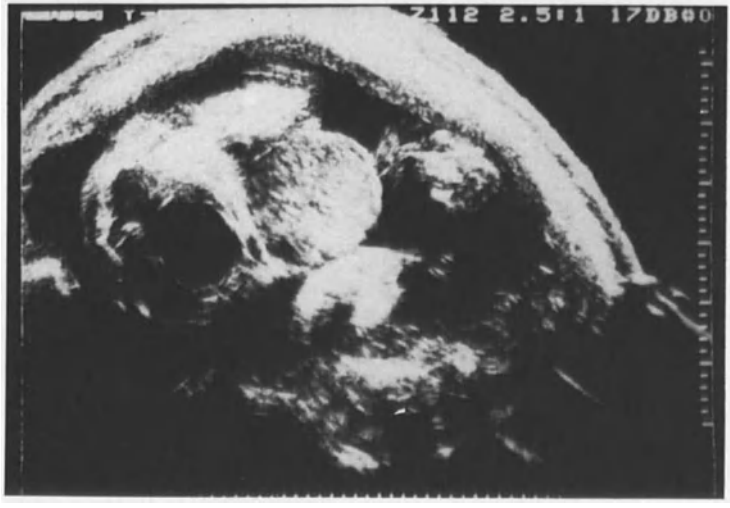

Fig. 12. Omphalocele: in front of the fetal abdomen the omphalocele, containing the fetal liver. Transverse scan

with the pediatric surgeon to plan the further management of the individual case (Figs. 11 and 12). As omphalocele is frequently combined with genetic disorders, amniocentesis should be performed in every case.

In cases of *gastroschisis* the bowel floats freely in the amniotic fluid. The defect in the abdominal wall can be exactly measured (Fig. 13).

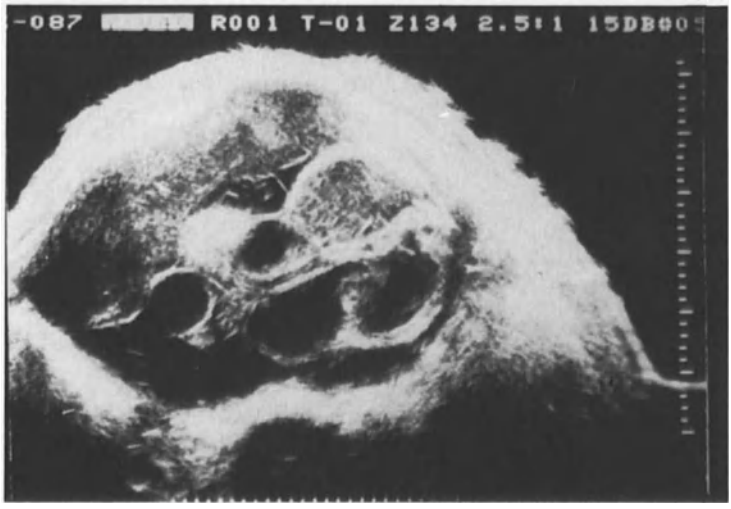

Fig. 13. Gastroschisis: the defect in the abdominal wall can be measured. Bowel freely floats in the amniotic fluid. Transverse scan

Malformations of the Urogenital Tract. The fetal kidneys are rather frequently affected by malformations. From the 18th week on the fetal kidney can be outlined and measured (Bernaschek and Kratochwil 1980; Garrett and Kossoff 1978).

Potter's syndrome is characterized by missing kidneys and bladder. Because of the agenesis of the fetal kidneys no amniotic fluid is present (Garrett and Kossoff 1978; Hansmann et al. 1979). Sometimes it may be difficult to decide whether the bladder is absent or was just previously emptied. In such cases the examination is prolonged to estimate the hourly fetal urine production.

Cystic lesions, either single or multiple, are easily detectable by ultrasonography (Reilly et al. 1979; Fig. 14). An infant with a unilateral severely damaged kidney is sent immediately after delivery to the pediatric surgeon for operation.

Urethral obstructions are characterized by a huge, overdistended bladder, hydronephrosis, and oligohydramnios (Fig. 15). A similar picture is found in prune-belly syndrome. It might be possible to relieve the obstruction by inserting a self-retaining catheter by percutaneous, ultrasonically guided puncture, thus performing a suprapubic cystostomy (Farrant 1980). However, this maneuver can result in polyuria, causing hydramnios and thereby inducing premature labor.

Posterior urethral valves have been correctly diagnosed prenatal (Kratochwil et al. 1972). Such cases formerly had a mortality of 50%. In our case the ureters were implanted into the skin and the valves resected transurethrally. After recovery of the kidneys, the ureters were reimplanted into the bladder.

Skeletal lesions. Since it has become possible to measure the long bones by ultrasonography (Queenan et al. 1980; Schlensker 1981), diagnostic interest has

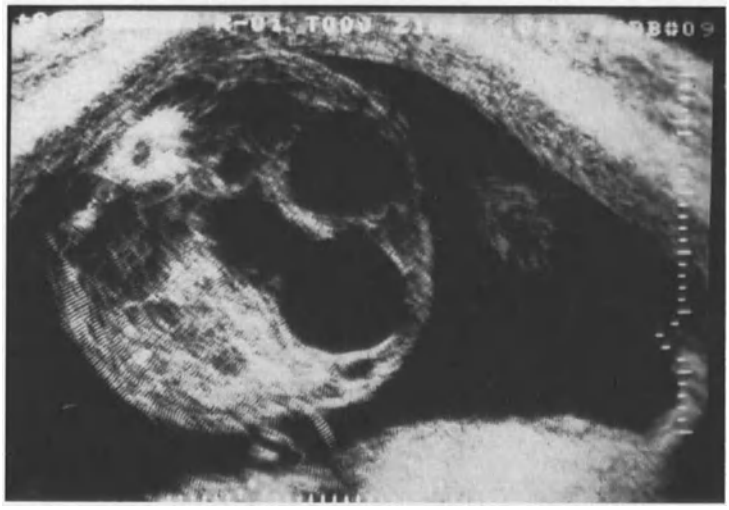

Fig. 14. Cystic kidney: fetus in cephalic position. The right kidney is normal; in the left kidney several cysts can be identified. Transverse scan

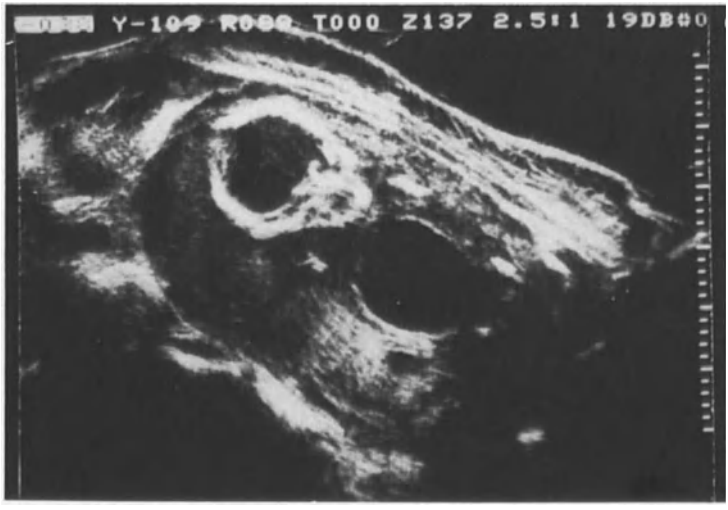

Fig. 15. Megalocystis: oligohydramnios; the fetus in breech presentation. The enlarged and overdistended bladder fills nearly the entire fetal abdomen

focused on the possible detection of skeletal anomalies. It has been successfully demonstrated that *osteogenesis imperfecta* and *aplasia of the long bones* can be detected by ultrasonography (Hobbins et al. 1979); however, a very experienced and patient examiner is required.

Teratomas of the fetal breech may cause considerable problems in the management of delivery if not detected antenatally (Gergely et al. 1980). These tumors

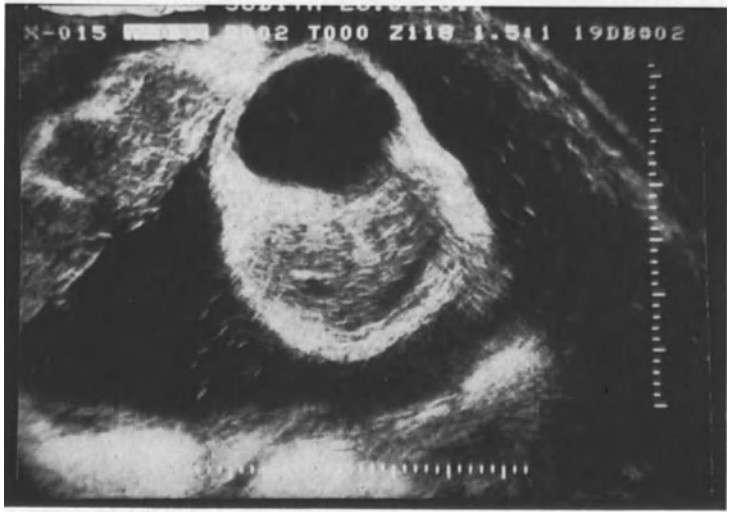

Fig. 16. Benign teratoma of the fetal breech. Besides solid masses, cystic portions due to degeneration can be seen

commonly appear solid in ultrasonography but, due to regression, may contain cystic portions as well (Fig. 16).

Conclusion

Ultrasonography can be a very powerful instrument in the hands of experienced examiners, but as prenatal diagnosis of fetal anomalies is only the first step in the management and counseling of the patient, close interdisciplinary cooperation is necessary.

As has been demonstrated, the prognosis of such anomalies could be considerably improved. Elective cesarean has benefitted infants with lesions causing dystocia.

Early notification of surgeons and neonatologists has reduced delays in postnatal evaluation and treatment. Considerable improvement has been achieved for babies with diaphragmatic hernia and meconium peritonitis, who develop respiratory insufficiency and may die immediately after birth (Canty et al. 1981; Toulonkian and Hobbins 1980). Further close cooperation will increase our experience in management and counseling to benefit our patients.

Summary

Ultrasonography is today able to detect many details of the fetal anatomy, so that prenatal diagnosis of malformations becomes possible. However, as 60% of all detected fetal abnormalities develop spontaneously, a screening program is

recommended. At least two ultrasonic examinations should be performed. To cope with the huge number of examinations obstetricians in their offices are integrated in this program. Women carrying fetuses with identified or suspected anomalies are sent for further evaluation, management, and counseling to special clinics.

The task is to differentiate between lesions which are incompatible with postnatal life, lesions which will necessitate a supporting person for the life of the patient, and lesions which are correctable by surgery. To gain more experience, close cooperation with neonatologists, pediatric surgeons, neurologists, and geneticists is recommended.

The first advantages of early prenatal diagnosis have been elective cesarean and reduction of postnatal evaluation and treatment required.

Résumé

Actuellement, l'échographie donne de nombreux détails de l'anatomie fœtale, de telle sorte qu'un diagnostic prénatal de malformations peut être établi. Un programme de screening est donc à établir pour faire une surveillance continuelle de ces anomalies fœtales. Deux examens aux ultra-sons sont nécessaires au minimum. Pour parvenir à un plus grand nombre d'examens, l'échographie devrait être considérée comme une aide à la naissance.

Il doit être établi qu'il faut faire une différence entre des lésions qui ne sont pas compatibles avec la vie, des lésions qui permettent une survie prolongée et des lésions qui sont correctibles chirurgicalement. Une collaboration étroite entre néonatologistes, chirurgiens pédiatres, neurologues et généticiens est nécessaire pour améliorer l'étude de ces anomalies.

Les premiers résultats du diagnostic prénatal sont de raccourcir les investigations post-natales, de façon à permettre un traitement précoce.

Zusammenfassung

Heutzutage vermag die Sonographie viele Details der fetalen Anatomie darzustellen, so daß eine pränatale Diagnostik von Fehlbildungen möglich ist. Da jedoch 60% aller entdeckten fetalen Anomalien einen kontinuierlichen Verlauf zeigen, ist ein Screeningprogramm vonnöten. Mindestens 2 Ultraschalluntersuchungen sind erforderlich. Um mit der großen Anzahl von Untersuchungen zurande zu kommen, werden die niedergelassenen Geburtshelfer in dieses Programm mit einbezogen. Sicher diagnostizierte und fragliche Anomalien werden zur weiteren Abklärung, Behandlung und Beratung an Spezialkliniken überwiesen.

Es wird die Aufgabe sein, eine Unterscheidung zu treffen zwischen Läsionen, die mit dem Leben unvereinbar sind, Läsionen, die lebenslange Betreuung erfordern und Läsionen, die chirurgisch korrigierbar sind. Eine enge Zusammenarbeit

zwischen Neonatologen, Kinderchirurgen, Neurologen und Genetikern ist nötig, um den Erfahrungsschatz zu vergrößern.

Die ersten Erfolge der pränatalen Diagnostik betreffen elektive Sectio caesarea, Verkürzung der postnatalen Untersuchungszeit und frühe Behandlung.

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Antenatal Ultrasound Diagnosis of Congenital Malformations of the Urinary Tract: Results and Criticism

C. A. DELL'AGNOLA[1], U. NICOLINI[2], G. PISTOR[3], and G. ALZEN[4]

From October 1979 to June 1983 220 malformations were diagnosed antenatally at our hospital; 55 (25%) were malformations of the urinary tract, 21 of them hydronephroses and 34 of them dysplasias.

The accuracy of antenatal diagnosis was 63.8% in 44 investigated cases. In 22% of the cases diagnosis could not be confirmed postnatally. In 14 of 20 cases (70%) pathologic conditions diagnosed clinically could be confirmed by ultrasonography during the first months of life (dell'Agnola et al. 1983).

Twenty infants had to undergo surgery. Prenatal ultrasonographic findings could be verified by postnatal ultrasonography, infusion pyelography, and micturition cyst urethrography.

Microscopically and electron microscopically the parenchyma was always inconspicuous, and scanning electron micrographs of the ureteric ostia always exhibited a regular cell surface.

There was no case of an- or oligohydramnios among the infants operated on. Neither complete atresia of the renal pelvis (dell'Agnola and Nicolini 1983) nor quiet kidney, nor preoperatively positive urine cultures were found. In agreement with ultrasonography, there were positive urographic findings in every case.

Only initially was the time of birth influenced by prenatal diagnosis, leading to preterm delivery in four cases (23%). Since 1981 we have changed our procedure, in so far as we await termination of pregnancy by spontaneous birth (dell'Agnola and Nicolini 1983).

Good morphological and functional results were obtained in 14 of 17 cases of subpelvic stenosis (82%). This was verified by follow-up for more than 24 months of four children, three of whom had bilateral hydronephrosis, a follow-up for more than 1 year of three children, and for more than 6 months of seven children.

Prenatal ultrasound diagnosis of malformations of the urinary tract enables us to recognize congenital uropathies prior to birth and to avoid complications (dell'Agnola et al. 1983). Our experiences confirm the possibility of early treatment with less risk (dell'Agnola et al. 1983; dell'Agnola and Nicolini 1983; Mayor et al. 1975).

Indications for intrauterine nephrostomy or cystostomy are given *only* if further morphological and biochemical criteria, such as an elevation of *N*-acetylglucosidase, are considered.

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Summary

Fifty-five congenital malformations of the urinary tract (25%) were among 220 antenatally diagnosed malformations. Accuracy of diagnosis was 63.8% (44 cases); 20 infants had to be operated on. Good morphological and functional results could be obtained in 82% of the cases. Prenatal diagnosis of congenital uropathies provides early treatment burdened with less risk.

Résumé

Cinquante-cinq malformations du tractus urinaire (25%) ont été diagnostiquées chez 220 malformations retrouvées par échographies anté-natales. La précision du diagnostic était de 63,8% (44 cas). Vingt enfants ont dû être opérés. Des résultats morphologiques et fonctionnels satisfaisants ont été obtenus dans 82% des cas. Le diagnostic prénatal des uropathies congénitales permet un traitement précoce qui peut être entrepris avec le moindre risque.

Zusammenfassung

Unter 220 pränatal diagnostizierten Fehlbildungen waren 55 Fehlbildungen des Harntraktes (25%). Die Treffsicherheit der pränatalen Diagnose lag bei 63,8% (44 Fälle); 20 Kinder mußten operiert werden. Gute morphologische und funktionelle Ergebnisse wurden bei 82% der operierten Fälle erreicht. Pränatale Diagnose von kongenitalen Uropathien ermöglicht frühzeitige und somit risikoärmere Behandlung.

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Obstructive Uropathies Diagnosed in Utero. The Postnatal Outcome – A study of 43 Cases

B. Fremond[1] and J. M. Babut[1]

The first publication concerning ultrasonic antenatal diagnosis of obstructive uropathies was by Garrett in 1975 [9]. Since then, advances in ultrasound have greatly increased the potential for the antenatal diagnosis of these malformations. Many important studies have contributed to the rapid expansion in the obstetric use of ultrasound in this field [10, 14, 15]. Apart from abnormalities in renal development – type I polycystic disease (Potter's classification), multicystic kidney, etc. – which produce irreversible states, the recognition in utero of obstructive uropathies at a stage where they are totally asymptomatic allows effective treatment to be carried out before the appearance of more serious lesions.

Subjects

Between 1980 and 1983, 43 cases of obstructive uropathies diagnosed antenatally were seen at the paediatric surgical unit at Rennes. They presented with anatomi-

Table 1. Distribution of 43 cases of obstructive uropathy diagnosed antenatally

	Sex	
	M	F
Upper urinary tract obstructions (23 cases)	17	6
– 22 ureteropelvic obstructions		
– 1 hydronephrosis associated with bifid ureter		
Ureterovesical obstructions (11 cases)	8	3
– 3 primary obstructive mega-ureters		
– 2 vesico-ureteric reflux		
– 1 single ectopic ureter		
– 5 ectopic ureteroceles		
Bladder outflow obstructions (5 cases)	5	0
– 3 posterior urethral valves		
– 2 bladder neck obstructions		
Prune-belly syndromes (4 cases)	3	1

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cal or functional (reflux) obstructions. Four prune-belly syndromes are also included in the series, as the pathogenesis of this condition is probably related to obstruction.

The distribution of cases is shown in Table 1; there is, as usual, male predominance, with 33 male patients and 10 female patients. The earliest diagnosis was made at 15 weeks' gestation, a prune-belly syndrome with severe oligohydramnios, megacystis, mega-ureters, abdominal distension and ascites. The latest was at 39 weeks (slight pelvic dilatation not seen again postnatally).

The diagnosis of ureteropelvic junction syndrome was made at an average of 35 weeks (Fig. 1), that of ureterovesical obstruction at an average of 30.9 weeks, and that of obstruction distal to the bladder at an average of 27.4 weeks.

Polyhydramnios was present on two occasions: one of the patients was female with a minor uropathy as part of an incomplete prune-belly syndrome, and the other was in a serious condition owing to bladder neck obstruction.

The unfavourable prognostic significance of oligohydramnios is epitomised by the five patients who presented with this condition and who showed severe

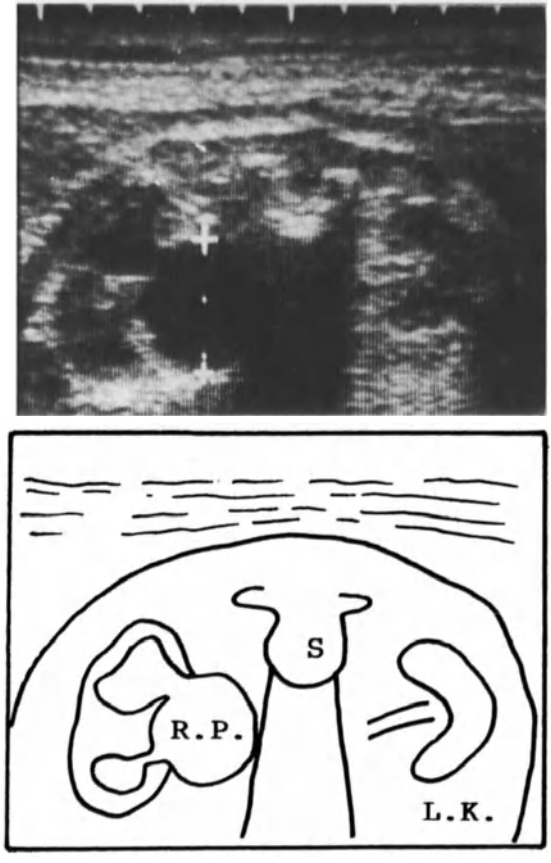

Fig. 1. Antenatal ultrasound at 37 weeks' gestation: transverse scan of abdomen. *S*, spine; *LK*, left kidney (normal); *RP*, right pelvis (dilatation)

obstruction; two died, of whom one survived for only a few minutes post-delivery (the uropathy had been diagnosed at 15 weeks).

With reference to antenatal treatment, in one case we attempted in utero decompression, but this failed owing to the position of the fetus

Preterm delivery for early relief of obstruction was carried out in 12 of the 43 cases: seven caesarean sections at 36–39 weeks' gestation, and five induced deliveries at 37–39 weeks' gestation. Six of these planned deliveries occurred away from our centre (Centre Hospital of Universitaire, Rennes), the infants being transferred to us after birth.

Results

The Neonatal Examination

1. Examination of Initial Status. Table 2 gives the results of the clinical examination, test of renal function, and urinary cytobacteriological examination, related to the type of uropathy present.

In only 18 of the 43 cases did a detailed *clinical examination* at birth (in the absence of antenatal information) reveal the existence of a uropathy (palpably

Table 2. Results of clinical examination, test of renal function and cytobacteriological examination of urine, related to type of uropathy

	Upper urinary tract obstructions	Obstructions of ureterovesical junction	Bladder outflow obstruction	Prune-belly syndrome	Total	
Clinical examination						
Normal	17	7	1		25	
Abnormal	6	4	4	4	18	
Renal function						
Normal	23	9	3	3	38	
Moderate failure		1	2		3	
Severe failure		1		1 (died immediately)	2	
Cytobacteriological examination of urine						
Negative	21	9	5	0	35	42
Positive	2	2	0	3	7	plus one immediate death

enlarged kidney or bladder, abnormal micturition or aplasia of abdominal musculature).

Renal function was evaluated by tests for urea and creatinine levels and, in 16 cases, with ^{99m}Tc DMSA uptake studies. With the exception of the patient who died several minutes after birth, only one infant presented immediately with serious renal failure, which caused its death at 1 month (this case showed ectopic ureterocele with significant vesicoureteric reflux both on that side and the contralateral side). Three other cases had renal failure with moderately raised urea and creatinine levels and DMSA uptake was significantly lowered – to 8.5% in the case of vesicoureteric reflux, and to 12% and 39% in the two cases of posterior urethral valves.

Cytobacteriological investigation of urine revealed that urinary tract infections presented with early pyuria and bacteriuria in seven cases. The causative organism was *Escherichia coli* in six cases and *Klebsiella* in one. They were all successfully treated with antibiotics. On three occasions the infection was precipitated by urological investigation, but it developed spontaneously in the other four cases.

2. *Investigations with Regard to Aetiology.* Ultrasound was always the first investigation. In two-thirds of the cases the antenatal diagnosis was confirmed by postnatal ultrasound. The key diagnostic examinations were IVU and cystography, either suprapubic or retrograde. Because of difficulties in interpretation IVU was not carried out before 48 h. When renal failure is demonstrated in early neonatal life, IVU is unhelpful and may be poorly tolerated. Dynamic scan with ^{99m}Tc DTPA, combined with frusemide-induced diuresis, was performed in seven patients in the neonatal period. Cystoscopy was performed in 16 patients prior to the operation.

In two cases of *ureteropelvic obstruction,* the postnatal ultrasound examination gave false reassurance, suggesting diminution or disappearance of the antenatal findings; in these two cases second studies at 2.5 and 5 months showed frank hydronephrosis, which would have benefited from earlier treatment. On two occasions regression of a mild hydronephrosis occurred spontaneously.

IVU allows a classification into four stages (I–IV), according to Cendron, and was done in 20 patients in the neonatal period, allowing forty kidneys to be evaluated (Fig. 2).

In one case the IVU was normal, the hydronephrosis diagnosed antenatally having disappeared, and in four cases the obstruction was bilateral. There were eight cases of stage I, four cases of stage II, six cases of stage III, and five cases of stage IV, making a total of 23 abnormal ureteropelvic junctions.

In two patients retrograde cystography showed associated reflux, which was bilateral in one case.

Primary obstructive mega-ureter presented in a bilateral form in the three cases, with extreme dilatation in two.

In the cases of *duplex ureters with ectopic ureteroceles,* IVU showed the contralateral kidney to be normal in four of the patients and non-functional in one. The affected kidney was non-functional in two cases, and in the other three cases the

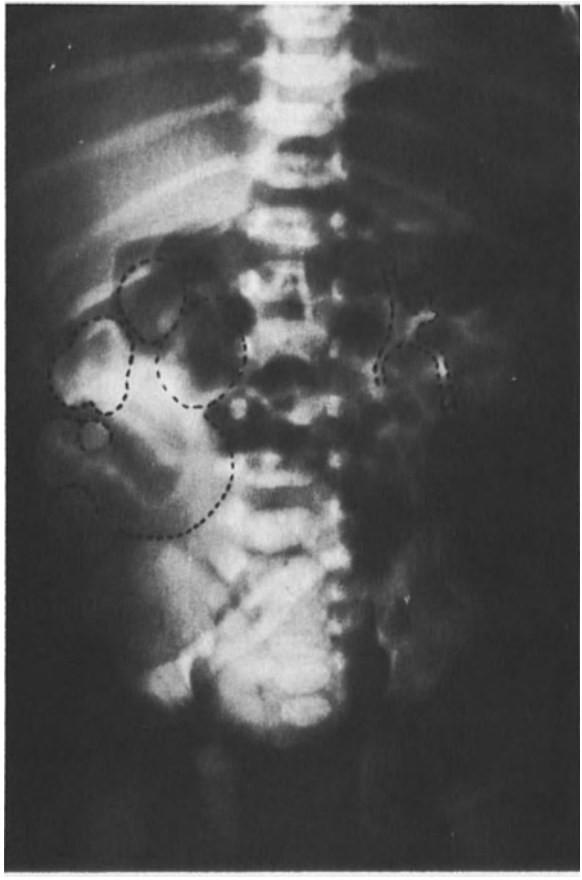

Fig. 2. Same case as in Fig. 1. IVU on 3rd day post-delivery: right ureteropelvic obstruction, stage III

ipsilateral lower pole was well visualised without pelvicalyceal dilatation. Only once could a faint opacification be seen in the upper pole.

In three cases of *bladder outflow obstruction* the IVU showed the typical findings: gross bilateral and symmetrical ureterohydronephrosis and tortuous ureters, with a hypertrophied bladder. There were three non-functioning kidneys in the other two cases. Suprapubic cystography revealed the cause to be posterior urethral valves (three cases) and bladder neck obstructions (two cases). In the cases with urethral valves, one case showed no reflux, but the other two showed gross unilateral reflux; one of the latter had associated severe renal dysplasia.

Among the three surviving patients with *prune-belly syndrome,* there was one minor form in a female patient, and two major forms.

Associated Malformations

These were uncommon. Apart from the four cases of prune-belly syndrome, of which only two were complete, there were one agenesis of the nucleus of the third

cranial nerve, associated with a ureterocele, and one large patent ductus arteriosus associated with a bladder neck obstruction.

Neonatal Mortality

There were three deaths. One of the patients with *prune-belly syndrome* died several minutes after delivery of pulmonary hypoplasia associated with severe oligohydramnios. One patient with *bladder neck obstruction* died at 7 weeks of cardiopulmonary failure secondary to severe sepsis, and a patient with *ectopic ureterocele* died at 1 month of renal failure.

Postnatal Management

The 42 surviving infants were divided into three groups (Table 3). Group 1 includes patients who did not require operations but who were treated with antimicrobial chemotherapy and followed up (12 cases, i.e. 28%). Group 2 includes patients who underwent surgery after some months of observation or medical treatment (8 cases, i.e. 20%). Group 3 includes patients who underwent surgery within the first 6 weeks of life (22 cases, i.e. 52%).

The seven cases of *ureteropelvic obstruction* in group 1 were all stage I. Group 2 included both stage II and III cases, the latest surgery being performed at 7 months. Group 3 contained all the stage IV cases, as well as a stage I case operated on early because of urinary infection. Anderson Hynes pyeloplasty, with a temporary nephrostomy and intubation of the anastomosis, was the procedure used in all cases. Only one nephrectomy was performed in the neonatal period for a stage IV case, and in retrospect this might have been avoided.

Only one child with *primary obstructive mega-ureters* was operated on in the neonatal period; cutaneous ureterostomy was performed for major obstruction.

Table 3. Distribution of uropathies among 42 surviving infants

	Group		
	1	2	3
Ureteropelvic obstruction	7	4	11
Hydronephrosis associated with bifid ureter	1		
Primary obstructive mega-ureter	1	1	1
Reflux	1		1
Single ectopic ureter		1	
Ureterocele		2	3
Posterior urethral valves			3
Bladder neck obstruction			2
Prune-belly syndrome	2		1

In two cases, urinary antisepsis was maintained and follow-up continued; one of these patients underwent surgery at 9 months owing to a secondary deterioration.

One of the two patients with *reflux* had a bilateral stage IV condition with renal failure, and underwent bilateral cutaneous ureterostomy, followed by ureteral remodelling and reimplantation on one side and nephro-ureterectomy on the other, but renal function remained poor. The other case had unilateral stage II reflux, which resolved spontaneously in 6 months with urinary antisepsis.

All five patients with *ureteroceles* underwent upper pole nephro-ureterectomy, three in the neonatal period and two at 3 months. Early intervention was elected when there was dilatation of the lower calyx on the affected side or changes in the contralateral kidney.

The cases of *bladder outflow obstruction* were all in group 3. Among those patients with posterior urethral valves, two required only endoscopic fulguration, while the third, who from the outset presented with severe renal failure, benefited from initial bilateral cutaneous ureterostomy. One of the patients with bladder neck obstruction benefited from early complete reconstruction using the Hendren technique with bladder neck YV-plasty. The other case could only tolerate cutaneous ureterostomy.

One of the three patients with *prune-belly syndrome* underwent bilateral ureteric diversion because of severe uncontrollable pyuria. The other two were merely observed.

Renal Pathology

For *ureteropelvic obstructions,* six renal biopsies and a nephrectomy were performed in three cases of stage III, and in four cases of stage IV hydronephrosis. The biopsy was normal in the three stage III patients. In two of the stage IV patients, biopsy showed only lesions of severe hydronephrosis, while the other two cases showed microcystic changes associated in one case with typical dysplastic lesions (primitive ducts surrounded by a collar of mesenchyma).

In patients with *ureterovesical obstructions,* dysplastic anomalies were found particularly in association with ectopic ureters, which correlates well with the theory of Mackie and Stephens [19]. In the patient with a single ectopic ureter entering the bladder neck, associated with gross mega-ureter and renal atrophy, the dysplasia was severe. Histological examination revealed numerous islets of cartilage.

In four of the five patients with ectopic ureteroceles with duplex systems, the upper pole was dysplastic and microcystic. Biopsy of the retained lower pole was performed twice, and this pole found to be equally dysplastic. The contralateral kidney was also dysplastic in one case. A renal biopsy conducted in one of the patients with primary obstructive mega-ureter was found to be normal; this was also the case in a patient with stage IV reflux.

Histological studies in three of the five cases of *bladder outflow obstruction* revealed dysplastic changes.

Outcome and Results

Among the 40 surviving cases the follow-up time was more than 1 year in 31, with a maximum of 4 years. The mean follow-up was 27 months. Outcome is based on examination of clinical (principally growth) and paraclinical states – ultrasound and radiological examinations, renal function tests and cytobacteriological studies of urine. Renal function was determined by urea and creatinine levels and by ^{99m}Tc DMSA uptake. The results are analysed in comparison with the various types of uropathy.

Twenty children with *ureteropelvic obstructions* were regularly checked and their heights and weights were normal (even in the bilateral cases). All the patients had sterile urine (without using urinary antimicrobial chemotherapy) and normal urea and creatinine levels. Of five patients with stage IV disease, three had post-

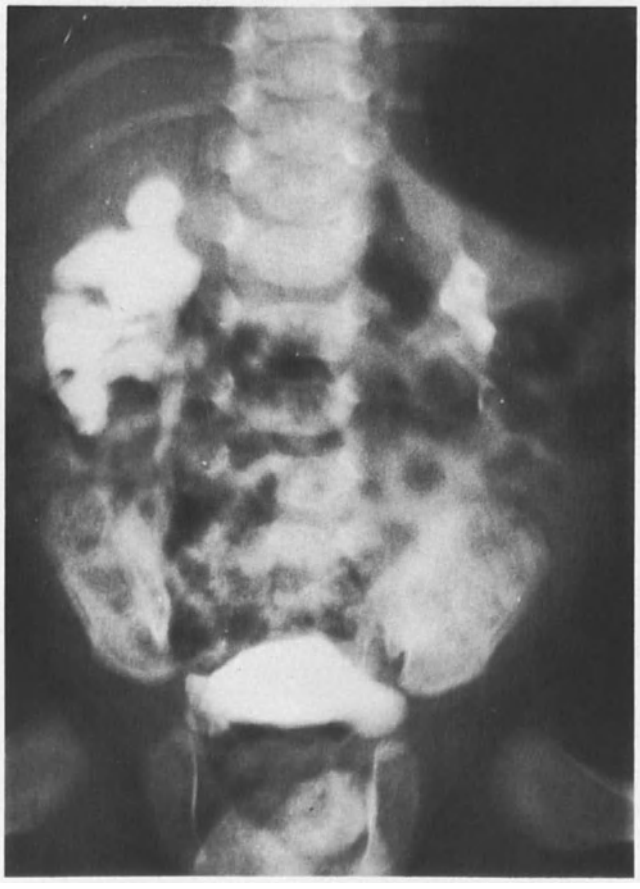

Fig. 3. Same infant as in Figs. 1 and 2, operated on at the age of 15 days. IVU 2 months later already shows an excellent recovery

operative DMSA uptake studies. The first two showed very low uptake levels, while the third showed good recovery, with an uptake of 68%. Ultrasound investigation was normal in all stage I cases, with the exception of one patient who had to be operated on at 3.5 years because of slowly, progressive deterioration. In all the surgical cases, postoperative ultrasound showed significant improvement but a discrete residual dilatation remained in the stage II and III cases, except in one stage III case where ultrasound returned to normal. In the stage IV cases, the residual dilatation was more marked, and in one case the hydronephrosis remained unchanged. IVU confirmed these results: excellent recovery in the stage II and III cases (Fig. 3), perfect stability in the stage I cases, and poor results in the stage IV cases, with two virtually non-functioning kidneys. One kidney secreted at 28 months, but with significant parenchymal atrophy. Of the two remaining stage IV cases, one underwent a nephrectomy in the neonatal period for a totally destroyed kidney, the other kidney showing a good functional recovery but with significant residual dilatation.

Of the three patients with *primary obstructive mega-ureter,* two underwent surgery, the first in the neonatal period for gross ureterohydronephrosis (bilateral cutaneous ureterostomy, followed by bilateral reimplantation and finally closure of the cutaneous ureterostomies at 20 months). The second patient underwent surgery at 9 months for a progressive dilatation of the pelvicalyceal system (tapering and reimplantation of both ureters). The third patient is under observation, having presented with bilateral gross ureteral dilatation, but with less severe

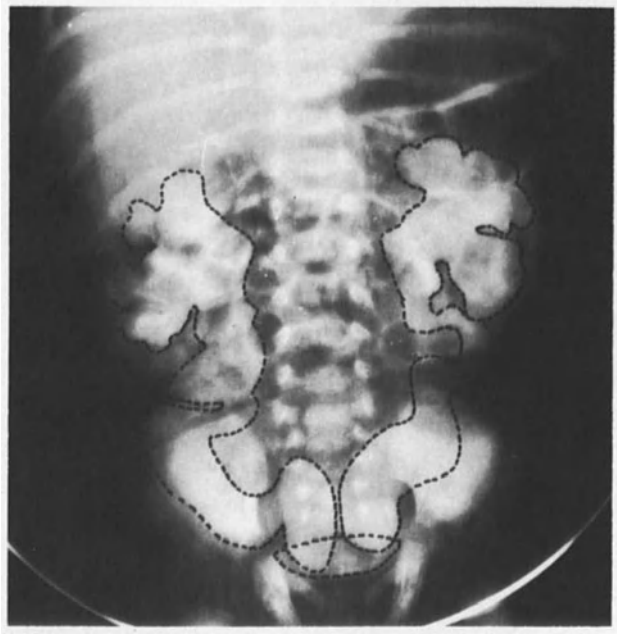

Fig. 4. Primary obstructive mega-ureter. IVU on the 3rd day after birth: renal function is normal. Straightforward follow-up maintaining urinary antisepsis

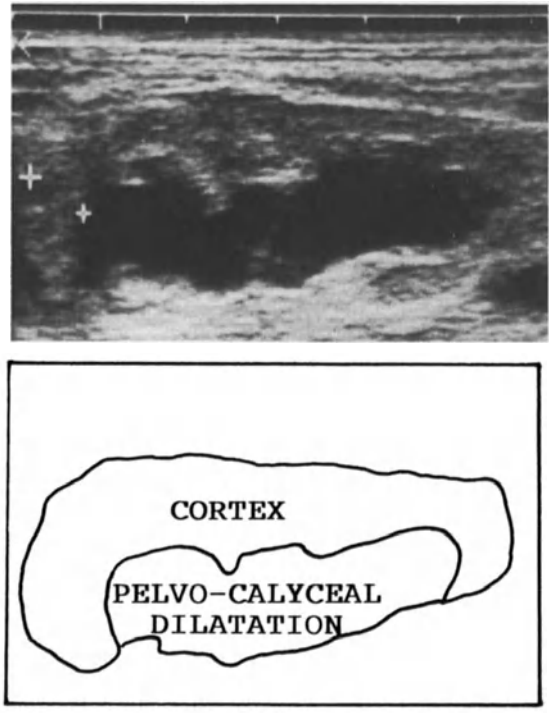

Fig. 5. Same case as in Fig. 4. Ultrasonic sagittal scan of the right kidney at birth: the pelvicalyceal system is dilated, but the cortex is of normal thickness

pelvicalyceal dilatation (Figs. 4 and 5). The IVU at 9 months showed a significant improvement (Fig. 6), which was confirmed on ultrasound scan (Fig. 7). In the first two cases the results are very satisfactory, with normal growth, normal renal function and constantly sterile urine (without urinary antimicrobial chemotherapy).

Of the five patients with *ectopic ureteroceles,* one died of renal failure at the age of one month. This patient presented with gross contralateral retention and severe bilateral renal dysplasia. The surviving four patients have very satisfactory results after upper pole nephro-ureterectomy without excision of the ureterocele itself, and show normal growth, constantly sterile urine, and normal renal function. The IVUs all show that the contralateral kidneys and ureters are normal and the lower poles exhibit good function without pelvicalyceal dilatation (even in one case in which the lower pole was initially non-functioning, with no DMSA uptake). In one case secondary contralateral stage II reflux appeared which on cystoscopy showed no abnormality of implantation. This child has remained well with antimicrobial therapy. Radiological and cytoscopic investigations confirmed good collapse of the ureterocele in all cases.

Among the five patients with *bladder outflow obstructions,* there was only one death, from hyaline membrane disease and septicaemia at 7 weeks. The remaining four patients showed varying degrees of severity: all have normal growth, with sterile urine and normal urea and creatinine levels. The surviving patient with bladder neck obstruction with gross upper urinary tract dilatation was treated with

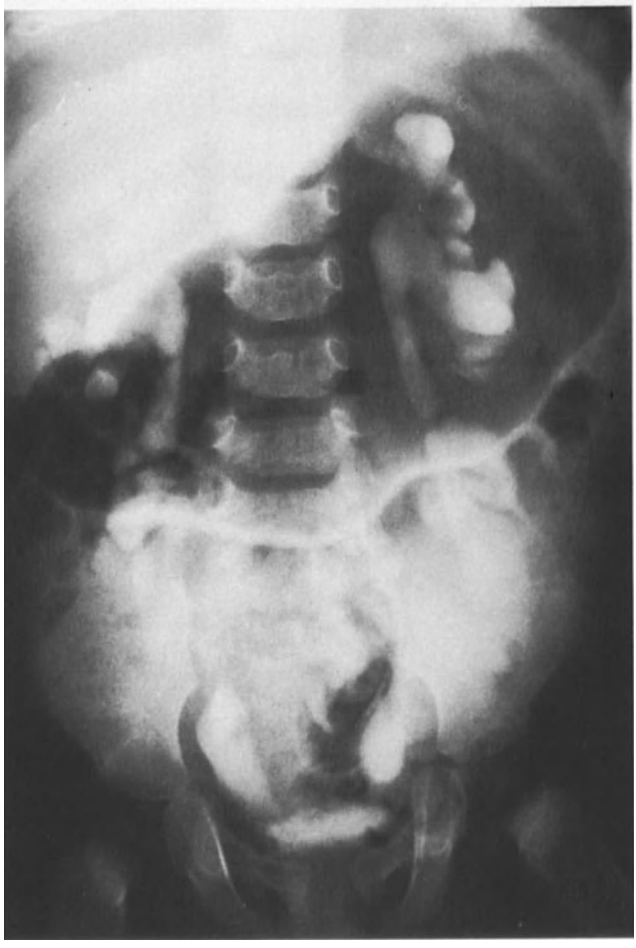

Fig. 6. Same case as in Fig. 4: IVU at 9 months shows a good spontaneous improvement

bilateral ureterovesical reimplantation, ureteral tapering, and bladder neck YV plasty at the age of 6 weeks, after initial suprapubic drainage. The outcome with regard to function and continence is excellent 4 years later, although with mild residual dilatation. Regarding the three patients with posterior urethral valves, one underwent a bilateral cutaneous ureterostomy at birth because of respiratory distress and precarious renal function, followed by endoscopic fulguration at 7 months, bilateral ureterovesical reimplantation at 1 year, and finally by closure of the cutaneous ureterostomies. This patient had a total DMSA uptake of 39% at birth, which increased to 88% at 3 years. Significant residual dilatation persists, but functional recovery and continence are excellent. The other two patients simply underwent endoscopic fulguration with a good result in one case, but it is too early to give a prognosis in the other, as one of the kidneys is totally dysplastic and non-functional, and the other shows a DMSA uptake of 24% at 6 months.

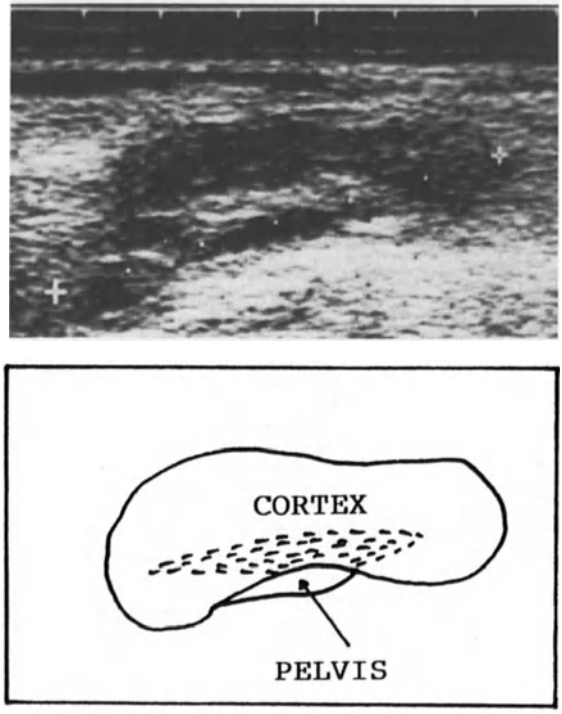

Fig. 7. Same case as in Fig. 4: ultrasonic sagittal scan of the right kidney at 9 months. The picture is almost normal, with good cortico-medullary differentiation

In the three cases with sufficient follow-up the outcome is satisfactory, given the initial overall severity of the condition, demonstrating that neonatal relief of obstruction allows significant recovery.

One of the patients with *prune-belly syndrome* died shortly after birth, and the surviving three showed conditions of varying severity. The first case was a female patient who presented with hypoplasia of the anterior abdominal wall, moderate dilatation of the pelves, and atonic ureters; this resolved on its own, marred only by an early urinary tract infection, and IVU at 3 months was normal.

The other two patients had classic forms. One had a major form with urachal fistula but without obstruction of the urethra; the other had a less severe form without obstruction, but was treated in the neonatal period for severe urinary tract infection which was not controllable with antibiotics. Both underwent bilateral cutaneous ureterostomy. These two children show normal growth but permanent pyuria despite antimicrobial chemotherapy, owing to the cutaneous ureterostomy. Follow-up has not been long enough to predict how renal function will develop (7 and 15 months).

Discussion

The fetal kidneys are derived from the metanephros, which is formed by the interaction of the ureteric bud with the nephrogenic blastema after the 5th gesta-

tional week. Urine secretion begins very early, in the 6th week [18], and nephrogenesis continues until the 36th week [25], while renal maturation continues for several years after birth. Any obstructions of the urinary tract during embryogenesis will inevitably reflect back on to the kidney.

The kidney may be visualised with ultrasound from the 15th week [17]. From the 20th week, the outline and the structure of the kidneys can be seen, and the shape of the pelvis and calyces can be assessed. The ureters are normally invisible; the maximum diameter of the bladder should not exceed 50 mm at term delivery. The echoanatomy of the normal fetal urinary tract is now sufficiently well known [2, 11, 12] to allow the discovery of an increasing number of abnormalities.

Much less is known on the other hand about renal function in the fetus. Calculation of urine output is feasible [7], possibly after administration of frusemide to the mother [32]. The best reflection of major renal failure is the presence of early oligohydramnios, particularly prior to the 20th week [15]. In our series, two of the patients presenting with oligohydramnios died. Polyhydramnios may result from the polyuria arising from the inability of the kidneys to excrete a concentrated urine, or is sometimes secondary to intestinal obstruction secondary to a grossly dilated urinary tract. The pejorative feature of hydramnios is not seen in this series, where it was only noted twice, once with a minor uropathy. At the present time it remains impossible to evaluate renal function above an obstruction and to assess its potential to recover.

The pathophysiology of in utero urinary tract obstructions is still inaccurate; only certain lesions produce the mechanical lesions of hydronephrosis, whereas others are followed by irreversible damage to renal development, producing cysts and dysplastic lesions. It has been well known since the work of Beck [3] on foetal lambs that obstructions developing in the first half of pregnancy cause irreversible changes, and clinical experience confirms this [4]. In our series it was among the cases with the earliest ultrasonographic detection that these lesions were observed. We have also noted that the most distal obstructions were diagnosed earliest. Another theory, parallel to that of Beck, stresses the relevance of the ectopic bud in the pathogenesis of cysts and dysplastic lesions [19].

After birth, these renal lesions may be considerably aggravated by infection, and may culminate in the triad of hydronephrosis, pyelonephritis and dysplasia. Functional impairment depends on the number of nephrons destroyed or damaged and begins with impairment of tubular concentrating power, while glomerular filtration may be totally conserved. Later, in the absence of treatment, the decreasing number of nephrons leads to renal failure if the pathology is bilateral.

Antenatal diagnosis of obstructive uropathies permits effective treatment for two features of the triad, namely hydronephrosis and pyelonephritis. Dysplasia is irreversible: in the cases of serious obstructions with severe oligohydramnios occurring at less than 20 weeks' gestation, ultrasound detection occurs too late [4], and at this stage attempts at urinary diversion in utero are not of proven efficacy. In the case of less severe obstructions with moderate or no dysplasia, we do not know whether drainage of urine before term (either by in utero amniotic shunting or after early delivery) protects, or at least limits the damage done to the nephrons.

Does urinary tract obstruction significantly alter renal function in the last months of intrauterine life, and is this hypothetical damage irreversible and unsusceptible to immediate postnatal treatment at term [31]? Interest in utero drainage has tended to decline, after an initial phase of enthusiasm. Would certain forms of urethral valvular obstruction benefit from a vesico-amniotic shunt? This has as yet not been clearly demonstrated. In cases of bilateral hydronephrosis which are due to ureteropelvic obstructions, early postnatal intervention obviously seems less dangerous than in utero drainage [27]. Eventually, in utero diagnostic intervention by percutaneous needle placement (to assess urinary concentrations, to study the reaction of diuresis to frusemide, or to inject contrast medium) may be of value in the most severe cases, where prognosis after birth may be compromised by hypoplastic pulmonary disease secondary to oligohydramnios. The main function therefore of antenatal diagnosis is, in the vast majority of cases, to give an opportunity to act immediately after birth. The prevention of urinary tract infections which cause secondary lesions of pyelonephritis, and the early relief of obstruction, are the best measures for the conservation or improvement of renal function. In contrast to other series of infants with renal malformations diagnosed postpartum [26], we have noted in this series the relative rarity of urinary tract infections.

Mathieu [20] has successfully shown the potential for recovery of renal function after relief of obstruction. The level of recovery is determined by the number of residual nephrons. Finally, recovery may be augmented by a compensatory hypertrophy, but this may be inhibited by the opposite kidney in cases of unilateral or predominantly unilateral uropathies. As we have observed several times in this series, DMSA uptake may be considerably improved after relief of obstruction, and an initially very low uptake does not necessarily indicate complete renal destruction. Thus, a pessimistic outlook based on a single DMSA scan is not justified [8].

Mechanical lesions (distension of pelvicalyceal system) are theoretically reversible, but many clinical and experimental studies have shown that the potential for recovery is limited proportionally with increased degree of obstruction and with the duration of the obstruction. Those obstructive uropathies that are discovered antenatally frequently show these two factors, posterior urethral valves being the best example. In addition, a more favorable outcome is obtained when the obstruction is proximal, particularly in ureteropelvic obstruction.

Long-term outcome in renal function is determined by two important prognostic factors – the degree of initial pathology, and age at first treatment:

1. It is quite obvious and largely proven that the greater the severity of the initial pathology, the greater is the risk of progression to end-stage renal disease [22, 30].
2. Mayor et al. [21] have shown that children with renal failure treated before the age of 1 year have in general a favorable long-term outcome, while those treated between 1 and 2 years of age are merely stabilised. Those treated after the age of 2 years deteriorate, generally after several years, even in the ab-

sence of urinary tract infection or of a recurrence of the obstruction. Puberty seems to precipitate this decline. The relationship between growth and puberty on one hand and the progressive deterioration of renal function on the other remains to be determined. McCrory [22] gives a more pessimistic outlook, stating that after the age of 6 months, recovery of renal function does not seem to be possible if it has been initially impaired.

Retrospective studies of children who have reached end-stage renal disease often show a prolonged delay between diagnosis and treatment of obstructive uropathies on the one hand, and the average age of first haemodialysis on the other [29, 16, 13], which is about 12 years. These series include a significant percentage (averaging 40%) of infants treated before they were 1 year old.

There is still, therefore, uncertainty about the potential improvement achievable by corrective surgery, and a follow-up period of at least 10 years is required to analyse the long-term results [30].

From these preliminary data it is easy to deduce two vital points in the treatment of antenatally diagnosed obstructive uropathies: prevention of urinary tract infection and early relief of obstruction.

One question that may be asked is, Is it in fact necessary to operate in all these cases in the first few days of life? Obviously this depends on the severity of the obstruction, and sometimes on its effect. In some cases one may indeed observe spontaneous regression of certain small hydronephroses. This occurred once in our series and the possibility has been emphasised by other authors [15, 5]. Are these therefore resolutions of pathological images or an aspect of normal physiology? One may add that small stage I junctional syndromes are on the whole well tolerated and stable, requiring not surgery, but close ultrasound observation. Of the nine patients with stage I junctional syndromes in our series two underwent operations, one in the neonatal period because of an early urinary tract infection, the other at the age of 3.5 years owing to as slowly progressive dilatation of the pelvicalyceal system. The latter case demonstrates the necessity of long-term follow-up over a number of years. Lastly, the primary obstructive mega-ureters often show a spontaneous regression after birth [6, 23], and this occurred once in our series. Only the major forms may eventually require early surgical correction or diversion.

In contrast, it is necessary to be particularly vigilant when faced with reassuring neonatal investigations which show a regression of the antenatal views. Our series includes two such cases. Ultrasound follow-up must therefore be used to screen for the reappearance of antenatal images.

The obstruction must be relieved in the neonatal period by the following methods:

1. For posterior urethral valves, the commonest procedure is no longer cutaneous diversion but simple valvular removal, leaving the treatment of upper urinary tract lesions until later [1, 24].
2. For the ureteropelvic obstructions beyond stage II, as shown by the good results we obtained in stages II and III, surgery must be performed in the

neonatal period. In stage IV the prognosis remains poor despite early intervention, as we have seen above (only one good result in five stage IV cases). Even so, the treatment must be conservative.

3. For ectopic ureteroceles, the upper pole partial nephro-ureterectomy combined with ureterocele collapse [28] gave good results.

Conclusions

This study clearly shows that antenatal diagnosis allowing early postnatal surgery is a major contribution to improvement of the prognosis for obstructive uropathies. For several years the number of children admitted with severe urinary tract infection, septicaemia and renal failure has steadily declined. The expansion in the use of ultrasound in obstetrics will diminish the number of these severe cases with a poor prognosis.

As far as the long-term outcome is concerned, it is still too early to judge the results, and we have already seen that a minimum of 10 years is required. Nevertheless, the results obtained in this series allow us to be optimistic in the majority of these cases.

Summary

The ultrasonic antenatal diagnosis of obstructive uropathies is now common. This study of 43 cases in which the mean follow-up is more than 2 years already allows us to show the importance of early diagnosis. The importance of a thorough postnatal examination is emphasised as well as the importance of preventing urinary tract infection. Although relief of the obstruction in the neonatal period in many cases allows an excellent recovery due to the exceptional qualities of the urinary tract at this age, there remain serious uropathies associated with significant renal dysplasia, for which ultrasonic diagnosis in utero is still insufficient. The results obtained allow us to be optimistic regarding the prognosis for many of these children.

Résumé

Le diagnostic anté-natal échographique des uropathies obstructives est maintenant courant. L'analyse de 43 dossiers, dont le recul moyen est supérieur à 2 ans, permet déjà de démontrer l'intérêt du dépistage précoce pour le devenir post-natal. La nécessité d'un bilan complet post-natal immédiat est soulignée, ainsi que l'utilité de la prévention de l'infection urinaire. La levée de l'obstacle en période néo-natale permet dans bien des cas d'obtenir une récupération excellente, grâce aux facultés exceptionnelles de l'appareil urinaire à cet âge, mais il reste les uropathies graves avec dysplasie rénale importante, pour lesquelles le dépistage

échographique in utéro est encore insuffisant. Si le devenir à long terme est encore incertain, les résultats déjà obtenus permettent d'être actuellement optimiste pour l'avenir de beaucoup de ces enfants.

Zusammenfassung

Die pränatale Ultraschalldiagnose obstruktiver Harnleiden ist heute durchaus üblich. Diese Studie über 43 Fälle, bei denen die Nachbehandlung im Durchschnitt 2 Jahre dauert, zeigt schon jetzt, wie wichtig eine Frühdiagnose ist. Die Betonung liegt sowohl auf einer gründlichen postnatalen Untersuchung als auch auf der Bedeutung der Prävention von Harnwegsinfekten. Obwohl in vielen Fällen eine Behebung der Obstruktion in der neonatalen Periode eine ausgezeichnete Restitution aufgrund der außergewöhnlichen Eigenschaften des Harntrakts in diesem Alter zuläßt, bleiben doch noch ernsthafte Harnleiden einhergehend mit einer signifikanten Nierendysplasie. Hierfür ist die Ultraschalldiagnose in utero noch unzulänglich. Das Ergebnis dieser Studie erlaubt eine optimistische Prognose für viele dieser Kinder.

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Is Prenatal Ultrasonography of Any Advantage?

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Published reports on prenatally diagnosed malformations are becoming increasingly frequent. Both scientific literature and the press in general cover heroic feats – separation of Siamese twins, successful attempts at intrauterine surgery. Is medicine marked as biological artistry?

These or similar headlines may give hope to concerned parents, but they make us hesitate at the same time; attending doctors are often confronted with conflicts of conscience in deciding on abortion, delivery by planned cesarean section, or introduction of prenatal therapy.

Parents to be are often not satisfied with a mere description of the clinical findings. They want to hear and learn more about their unborn child. Is the life of their child expected to be loaded with burdens and sufferings? Will this child be accepted by and integrated into society? Will family life be severely influenced by the child's handicap? The attitude of the attending doctors has a significant impact on the decision of the parents. Motivation to seek abortion or to continue with pregnancy and to begin with carefully planned therapy depends in many cases strongly on the surgeon first consulted. According to our experience in Heidelberg, it is extremely difficult to relieve a pregnant woman's anxiety and concern and to obtain consent for therapy once these feelings have been introduced.

Critical questions request critical, but open answers. Besides professional qualification and a high ethical standard, an interdisciplinary consultation and the agreement of all specialists concerned are necessary before advice is given to the pregnant woman. The obstetrician who attends the mother-to-be knows best about her social environment; the pediatric surgeon has to recommend treatment and acts as a lawyer for the unborn; the specialist in human genetics advises on the origin and possible outcome of the condition; and the pediatrician specializing in neonatology has to care for the newborn, in some cases up to its premature death. Only by joint efforts, in an open discussion between parents and attending surgeons, can a way out of this labyrinth be found.

Success or failure of therapy is often influenced by maturity, by weight at birth, and especially by associated malformations. Unrecognized additional malformations at times pose severe problems.

Between 1975 and 1982 11,372 babies were delivered at the University of Heidelberg Department of Gynecology and Obstetrics. The rate of malformations

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was 1.9%, with a notable increase from 1980 on. Of all malformations, 95% were diagnosed by ultrasound, 91% at the university hospital itself. In 98 cases suspected malformation was diagnosed at other hospitals and confirmed by us. Thirteen malformations were diagnosed at other hospitals and the patients then referred to the Heidelberg center for further care. In a further ten cases, other examinations led to the diagnosis of suspected malformation.

Among 161 cases of severe malformations, lesions of the central nervous system constituted the major portion, at 42% ($n = 67$), followed by malformations of internal organs and the anterior abdominal wall ($n = 65$; 40%). Malformations of the extremities were seen in two cases (1%), and other rare malformations (27 cases) made up the remaining 17%.

In 32 cases malformations were not recognized during prenatal ultrasonography. Twenty-one of these patients had abnormal findings (anomalies of the amniotic fluid, abnormal fetal biometry, abnormal fetal movements); however, no diagnosis was made during ultrasound examination. The remaining 11 patients had completely normal findings with ultrasonography; in seven of these cases malformations of the cardiac system were detected in the newborn.

Because of the variety of the malformations observed, a classification into four groups is helpful:

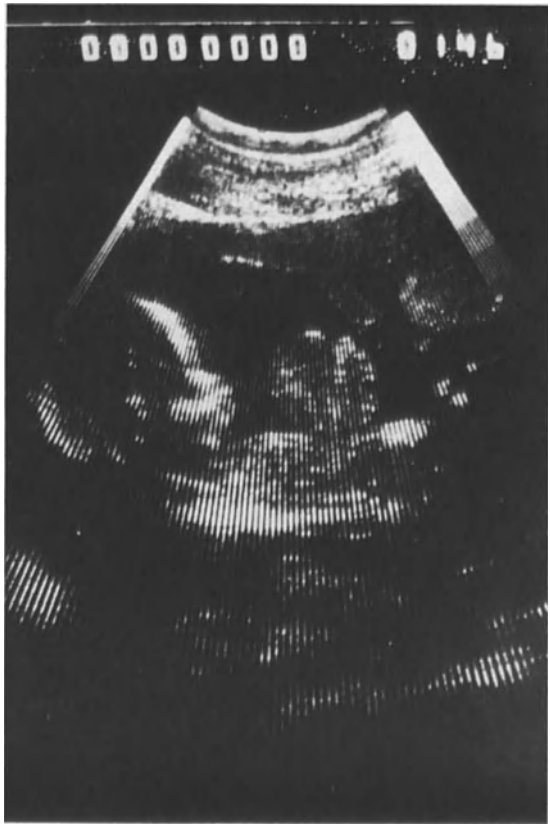
1. Single, completely correctable malformations
2. Insufficiently correctable malformations
3. Correctable malformations with additional, uncorrectable defects
4. Malformations incompatible with life

Single malformations belonging to group 1 (Fig. 1a, b) and a combination of multiple malformations of this group are manageable. We were able to correct successfully an omphalocele combined with an atresia of the esophagus.

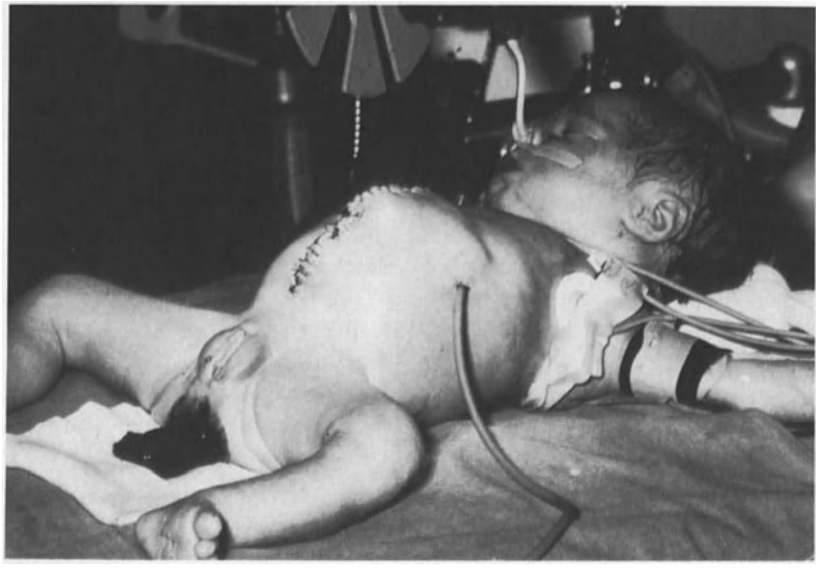
Group 2, with insufficiently correctable malformations, includes meningomyelocele with its special problems. The problems resulting from this malformation for parents and child are complex. A life of suffering begins, marked by countless consultations and hospital stays in the different medical specialties involved: urology, neurosurgery, orthopedics, pediatrics. The family, especially the mother, cannot but dedicate their life fully to the care of the child concerned. In this way other members of the family – brothers and sisters, the husband – are neglected. Problems evolve which are often hidden from our view. This sequence may arise from the decision to operate upon such a child. But how to act? We cannot and must not give recipes. Any help for a decision to find a way out of this dilemma has to be guided by high ethical responsibility and by joint counseling of the afflicted parents by the different medical specialists.

Group 3 – correctable malformations with additional, uncorrectable defects – includes trisomy 21, or Down's syndrome, which is often accompanied by organic heart defects, atresias of the gastrointestinal tract, malformations of the urinary system, and others.

To give an example of group 4 – malformations incompatible with life – I would like to mention the case of a fetus with a relatively minor omphalocele diag-

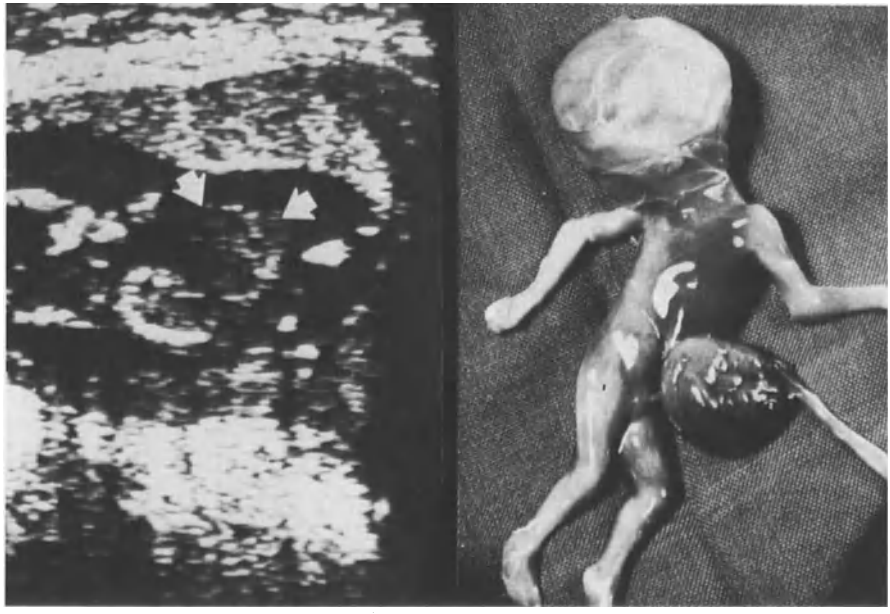


a



b

Fig. 1a, b. Omphalocele, Jones type III. **a** Twentieth week of pregnancy; **b** after surgical correction with amnion and mobilized skin



a b

Fig. 2. a Omphalocele, ultrasound at 17th week of pregnancy. Subsequent amniocentesis revealed trisomy 18. **b** Aborted fetus

nosed by sonography in week 17 of pregnancy. Amniocentesis, however, revealed the presence of trisomy 18 (Fig. 2a, b). The feasible correction of the omphalocele is by no means decisive for the further life of the child. These children are in need of our special care as the expression of our deep respect for life.

Apart from all these difficult, often only inadequately solved problems imposed on us by prenatal ultrasonography, a number of advantages resulting from early recognition of malformations in the expected baby are evident. Among these is planned delivery by cesarean section, which reduces the risk for the newborn. Risk factors such as the contamination of ruptured omphaloceles and gastroschisis associated with vaginal delivery can be eliminated. In a gastroschisis with concurrent liver prolapse, the risk of rupture of the liver is avoided by cesarean section. The expectant mother can be referred to a specialized and well-equipped center in time for delivery, thus eliminating the risk of expensive and time-consuming transport of the newborn by helicopter or ambulance. Furthermore, risks associated with prolonged transport, such as hypothermia, aspiration, and electrolyte imbalance are avoided. Other advantages are a well-planned, 'elective' surgery for the expected baby.

Introduction of therapy in utero is of growing interest. Possible applications include: intraperitoneal transfusion in cases of Rh incompatibility; the elimination of urinary tract obstructions; the easing of intracranial pressure in hydrocephaly (although questionable); direct administration of drugs in certain diseases (cardiac

arrhythmia, hypo- or hyperthyreosis); elimination of massive chylothorax, ascites, and other diseases.

We observed a case of massive chylothorax and lung obstruction during the 37th week of pregnancy. After puncture the lung expanded fully and the date of delivery could be awaited calmly. A further case of bilateral chylothorax was seen by one of us (W.M.); bilateral puncture was performed immediately postpartum and succeeded, with expansion of the lungs and survival of the child.

In conclusion, without disregarding all the advantages of prenatal ultrasonography, we must not close our eyes to the dilemma posed by knowing in advance. Not only the parents but also the surgeons involved are often drawn into severe inner conflicts from which there is no generally valid way out.

In spite of medical technology, doubt and uncertainty remain in all cases. A sword of Damocles in the form of unrecognized additional malformations hangs over the heads of all those involved, as these additional malformations may have a severe impact on an otherwise promising prognosis and may pose serious ethical problems for both the parents and the surgeons.

Forced continuation of pregnancy despite knowledge of the existence of severe lesions in the baby and risking daring surgery without perspective amounts to surgical tightrope-walking. We must recognize the frontiers of the medically feasible.

Summary

With the percentage of prenatally diagnosed malformations rising, there is a growing need for the contribution of the pediatric surgeon regarding prognostic and therapeutic aspects for the unborn child. An important risk factor for the validity of any statement is the possible presence of undetected, associated malformations. In spite of all advantages prenatal diagnosis offers regarding carefully planned therapy, etc., doubt and uncertainty always remain.

Résumé

Avec la quantité croissante de malformations diagnostiquées à la période prénatale, la contribution du chirurgien pédiatrique est devenue plus importante sur le plan de l'évocation du pronostic et des attitudes thérapeutiques.

La possibilité de malformations associées non diagnostiquées alourdit le caractère pathologique de l'état de l'enfant et le risque. De ce fait, il reste toujours un doute et une insécurité dans le diagnostic prénatal et la planification thérapeutique doit en tenir compte de façon impérative.

Zusammenfassung

Mit dem wachsenden Anteil von pränatal diagnostizierten Fehlbildungen ist ein zunehmender Beitrag von seiten der Kinderchirurgen hinsichtlich Prognose und therapeutischem Vorgehen nötig geworden.

Mögliches Vorliegen nichtdiagnostizierbarer begleitender Fehlbildungen belastet die Zuverlässigkeit jeglicher Aussage mit einem Risiko. Trotz aller Vorteile, die die pränatale Diagnostik hinsichtlich Therapieplanung etc. mit sich bringt, bleibt doch ein Rest von Zweifel und Unsicherheit.

Consequences of Antenatal Diagnosis for Pediatric Surgery

A. M. Holschneider[1], M. Baumgartner[2], and C. Mascott[1]

From 1979 to 1983, 638 newborns were referred immediately after birth to the Pediatric Surgical Hospital of the University of Munich and operated on as emergencies. Such emergencies increased by one-third during this period. Since the Munich Perinatal Study did not register a rising number of malformations, we must conclude that the increased number of referrals was due to a more exact and extended antenatal diagnosis, leading to admittance of risk pregnancies to obstetrical centers, which in turn sent malformed newborns to our hospital. This means that the pediatric surgeon is more and more concerned with problems of antenatal diagnosis of congenital malformations and consequences arising from them.

Here we describe our experiences with the care of pregnant women bearing malformed fetuses and discuss the state of the art of pre- and perinatal treatment.

Patients

The number of ultrasound examinations during pregnancy performed at the first University Gynecological of Munich doubled from 1968 to 1982. Whereas less than 6000 examinations were carried out per year in 1968 and 1969, the number exceeded 13,000 in 1982 (Fig. 1).

According to expectations, the number of malformations diagnosed rose during the same period; 23 malformations were found in 1979 and 78 in 1983. Malformations were diagnosed antenatally in a total of 229 fetuses from 1979 to 1983 (Fig. 2).

Whereas the ages of 1690 pregnant women with healthy fetuses ranged between 20 and 25 years, women with malformed fetuses were aged between 27 and 35 years (Fig. 3).

Kinds of Malformations

Table 1 classifies the malformations diagnosed at the ultrasound department of the First University Gynecological Clinic of Munich from 1979 to November 1983. Some malformations such as hydrocephalus, omphalocele, and hydronephrosis

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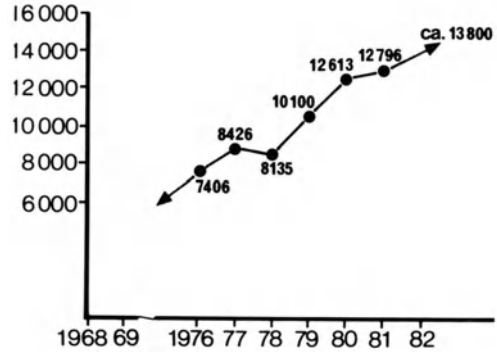

Fig. 1. Development of ultrasound diagnosis at the First University Gynecological Hospital of Munich, 1968–1982; number of examinations per year

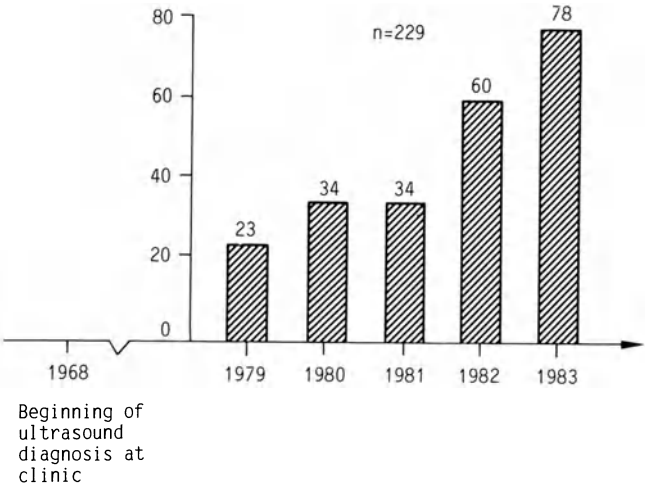

Fig. 2. Incidence of malformations diagnosed at the First University Gynecological Clinic of Munich, 1979–1983

could be diagnosed directly, while others, such as congenital heart disease or gastrointestinal anomalies, were detected indirectly by association with ascites, hydramnios, or cardiac dysrhythmia (Schäfer 1982).

Comparing the incidence and kinds of prenatally diagnosed malformations with the number and clinical signs of the newborns postnatally referred to us and operated on, one notes a preponderance of malformations of the central nervous system in both collectives, closely followed among our patients, however, by malformations of the gastrointestinal tract, the diaphragm, the abdominal wall, and

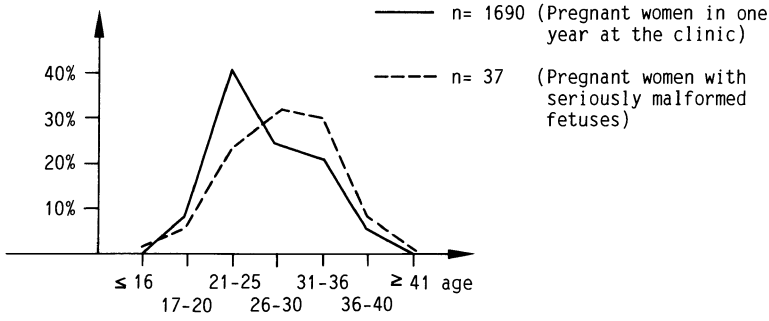

Fig. 3. Age distribution of pregnant women with malformed fetuses

Table 1. Malformations and diseases prenatally diagnosed at the First University Gynecological Hospital of Munich, 1979–1983

	1979	1980	1981	1982	1983	Total	(%)
Skull malformations	6	9	11	20	19	65	(28.4)
MMC + encephalocele	2	1	–	2	4	9	(3.9)
Cardiac anomalies	4	7	9	8	8	36	(15.7)
Esophageal atresia	–	–	1	2	–	3	(1.3)
Abdominal malformations	–	–	3	2	2	7	(3.1)
Omphalocele/gastroschisis	1	–	–	–	4	5	(2.2)
Renal malformations	–	3	6	5	20	34	(14.8)
Other anomalies[a]	4	7	1	7	7	26	(11.4)
Indirect signs: ascites, hydramnios	10	11	5	5	13	44	(19.2)
Total	27	38	36	51	77	229	(100)

MMC, Myelomeningocele

[a] Acardius, intracranial tumor, Siamese twins, hydrops, facial cleft, malformed extremities, thanatophoric dwarfism, coccygeal teratoma, etc.

the urinary tract. This discrepancy indicates that some malformations cannot be accurately diagnosed with antenatal ultrasound (Table 2).

Accuracy of Ultrasound Diagnosis

The uncertainty of antenatal diagnosis is shown by comparison of diagnoses on admission in pregnant women with malformed fetuses with ultrasonic or postnatal findings.

On the one hand, anencephalus, cardiac dysrhythmia, and hydrocephalus were correctly diagnosed in 50%–75% of cases: on the other, congenital heart

Table 2. Malformations treated neonatally at the Pediatric Surgical Clinic of the University Children's Hospital of Munich, 1979–1983

	1979	1980	1981	1982	1983	Total	(%)
HC	43	44	40	53	58	238	(36.9)
CNS–MMC	18	10	10	19	17	74	(11.5)
Diaphragm	6	10	12	9	8	45	(7.1)
Abdominal wall	7	9	15	6	15	52	(8.1)
Intestinal tract	32	31	38	39	38	178	(27.6)
Congenital malignant tumors	2	2	2	2	4	12	(1.9)
Urogenital tract	2	2	6	8	10	28	(4.3)
Others	2	2	3	5	5	17	(2.6)
Total	112	110	126	141	155	644	(100)

HC, Hydrocephalus; CNS, central nervous system; MMC, myelomeningocele

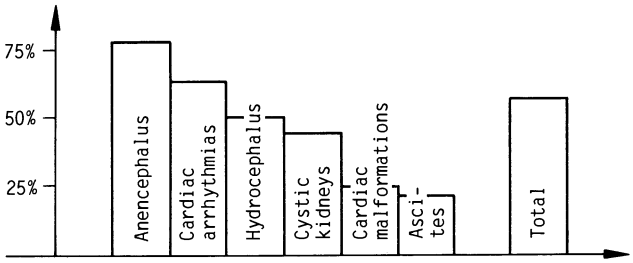

Fig. 4. Accordance of diagnoses on admission with ultrasound and postnatal diagnoses made at the First University Gynecological Hospital of Munich

disease, ascites, or even malformations of the gastrointestinal tract were seldom diagnosed (Fig. 4).

These figures are in accordance with our own experiences obtained in continuous care of 46 pregnant women who gave birth to malformed infants treated at our hospital post partum. Antenatal diagnosis was correct for 38 of these 46 infants (82.6%). However, diagnosis was incomplete for 18 infants (39.1%), since associated malformations such as esophageal atresia, bowel atresia, Hirschsprung's disease, and congenital heart disease were not recognized. Moreover, myelomeningocele in correctly diagnosed hydrocephalus, cloacal malformation in proven megaureter, and megalocystis were not diagnosed.

Occasionally a diagnosis remained unclear (10.8%) or was wrong (6.6%), for instance in an infant with hydramnios and a missing stomach who also had a diaphragmatic hernia and esophageal atresia, or in an infant with ultrasonographically diagnosed hydrocele testis in whom a ruptured abdominal neuroblastoma and a scrotum filled with blood were found postnatally. Other cases were a newborn with hydramnios and a massively distended stomach who had, besides suspected

Table 3. Pre- and postnatal diagnoses of 46 continuously followed newborns from various gynecological hospitals of upper Bavaria, referred to and operated on at the Pediatric Surgical Clinic of the University Children's Hospital of Munich

Diagnosis	*n*	(%)
Correct (incomplete)	38 (18)	(82.6; 39.1)
Indistinct	5	(10.8)
Wrong	3	(6.6)
Total	46	(100)

Table 4. Summary of correct and diagnoses made with ultrasound at the First University Gynecological Hospital of Munich in 1982

Malformations[a]	Cases (*n*)	Correct	False-positive	False-negative	
Anencephalus	12	12	0	0	
Hydrocephalus	11[b]	10	0	1	26 (41.2%)
Craniofacial synostosis	1	1	0	0	
Microcephalus	2	1	1	0	
Vertebral clefts	8[b]	3	2	3	
Intracranial tumor	1	1	0	0	
Cardiac malformations	11	5	3	3	
Ventral clefts	5	2	0	3	
Cystic hygroma colli	1	1	0	0	
Renal malformations	9[b]	5	2	1	
Intraabdominal tumor	1	1	0	0	
Double malformations	1	1	0	0	
Total	63[c]	43 (68.2%)	8 (12.7%)	11 (17.5%)	

[a] Gastrointestinal malformations were not diagnosed
[b] In one case the fetus was difficult to visualize
[c] In one case autopsy was not performed (1.6%)

duodenal atresia, anorectal atresia and congenital heart disease; a baby who presented ultrasonographically as particularly big and turned out postnatally to have Beckwith-Wiedemann's syndrome; and an infant with megalocystis in whom urethral stenosis was suspected, but associated anorectal atresia and congenital heart disease were not (Table 3). Thus, in this heterogeneous group of patients from different gynecological hospitals in upper Bavaria, correct and complete diagnosis was made of only 20 pregnant women (43.5%). However, diagnosis was

entirely wrong in only three cases (6.6%). Comparing this patient collective with the homogeneous patient group of the First University Gynecological Hospital of Munich treated in 1982, one sees that malformations of the skull were accurately diagnosed in 41.2%, but that vertebral clefts were as problematic as malformations of parenchymatous organs or the gastrointestinal tract, particularly in breech presentations. In total, fetal malformations were correctly diagnosed for 43 pregnant women of the patient group (68.2%), whereas false-positive judgements were made in eight cases (12.7%) and false-negative judgements in 11 cases (17.5%) (Table 4).

Consequences of Antenatal Diagnosis

Interruption of Pregnancy

The consequences of antenatally diagnosed malformations differ considerably from country to country. Comparative investigations in a Helsinki perinatal group of 3110 newborns and in a Munich patient collective of 6228 newborns in the frame work of the ARVO-YLPÖ Study (Riegel et al. 1984) showed that 6.9% of the infants born in 1980 and 1981 in the Helsinki area had malformations, whereas

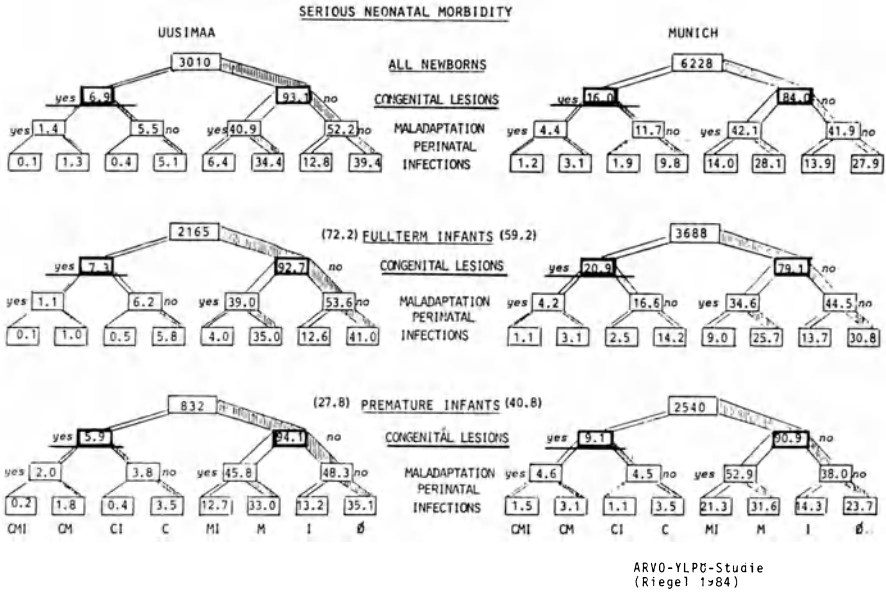

Fig. 5. ARVO-YLPÖ Study (Riegel 1984). Comparison of 3010 newborns from the Helsinki area with a Munich patient collective of 6228 newborns with regard to congenital malformations, maladaptation, and perinatal infections. Capital letters refer to the types of severe disturbance: *C,* congenital malformation; *M,* maladaptation; *I,* perinatal infection; *CMI,* congenital malformation + maladaptation + perinatal infection; *Ø,* none

Munich babies exhibited malformations 2.5 times as frequently (16%). Of babies born at term, 7.3% presented with malformations in the Helsinki area, whereas this was the case three times as frequently (20.9%) in the Munich region. In contrast, only slight differences appeared in prematures, with 7.9% and 9.1% respectively (Fig. 5). One can conclude that indications for interruption of pregnancy in the case of malformation were taken more liberally in Helsinki than in Munich.

According to West German criminal law (§ 218a, *Strafgesetzbuch* [StGB]), termination of pregnancy is allowed until the 22nd week post conceptionem or the 24th week post menstruationem if "pressing arguments indicate that the child will suffer from irreparable damage of health due to genetic or noxious antenatal influences so severe that continuance of pregnancy cannot be demanded of the pregnant woman." Since congenital malformations can often be diagnosed after that time only, interruption of pregnancy is possible in the F.R.G. only if the condition of the fetus is analogous to brain death, such as anencephalus or acardius, or if the malformation is incompatible with life, such as Edward's syndrome, Patau's syndrome or Potter's syndrome.

The physician should always keep in mind that in 1975 the Federal Court of Constitutional Law based its verdict on § 218a StGB on the principle that the fetus in utero is protected by the constitution as an independent legal entity, and that the state is required "to preserve and further growing life" (Krey 1976). This responsibility for protection is, as a matter of principle, also valid for the mother. However, the claim of the growing fetus to life is superior to the claim of the mother to free personal development and self-responsibility.

Psychological Aspects

We have experienced that parents of malformed fetuses show a positive attitude toward their malformed child which, however, underlies strong feelings of anxiety. Since there is intensive communication between the mother and the physician at each of the repeated ultrasound examinations, this anxiety can be reduced in stages, with the result that psychologically the mother handles the delivery of a malformed child much better than if she were surprised by the malformation after delivery. However, it is the task of the physician neither to persuade the parents to accept a severely malformed child nor to convince them of the usefulness of termination of pregnancy. It is most difficult to inform the parents objectively of the prognosis and the necessary postnatal therapeutic measures on the basis of an insecure antenatal diagnosis which cannot, as mentioned above, fully predict the extent and consequences of a malformation. Nevertheless, in every case all risks for the unborn child and long-term aspects for the family must be estimated to enable the parents to make a decision, also as far as postnatal therapy is concerned.

Therapy

At present, intrauterine treatment of congenital malformations is hardly possible. Although animal experiments of Haller et al. (1974, 1976) demonstrated promising aspects of intrauterine closure of diaphragmatic or abdominal wall defects in fetal lambs with diaphragmatic hernia or gastroschisis, and although Harrison et al. (1982a, b), Nakayama et al. (1983), Michjeda and Hodgen (1982) and Michjeda et al. (1983) showed the success of intrauterine ventriculoamniotic or vesicoamniotic shunts in rhesus monkeys, these experimental models cannot be transferred to human patients. Similar intrauterine procedures were performed on hydrocephalic fetuses by Jeanty and Rodesch (1981) and Clewell et al. (1982) and for malformations of the urinary tract by Kirkinen et al. (1982). However, these measures were not taken until the 26th–29th week of pregnancy, i.e., at a time when the value of intrauterine treatment is questionable. Above all, the main factor that speaks against intrauterine treatment at the moment is a lack of knowledge about pathophysiological principles, such as pressure in the fetal body cavities; about consistency of amniotic or cerebrospinal fluid, urine etc. during different stages of development; about the point in a pregnancy at which damage occurs and what treatment would be necessary. Also lacking is accurate diagnosis indicating whether an abnormality such as dilated ventricles represents a real pathologic condition, whether it may undergo reversal, or whether it will remain asymptomatic as arrested hydrocephalus.

Finally, the ethical question arises of whether a severely malformed child of a mother who can most probably give birth to further healthy children must – at considerable risk for both child and mother – be kept alive or not.

At the moment, there are only two viewpoints concerning treatment of an antenatally diagnosed malformation. The first is close control of the malformation to follow its development. Among our patients we experienced at least two different kinds of development of hydrocephalus: on the one hand, ventricular dilatation diagnosed as early as in the 17th–18th week of gestation but with only mild further enlargement later on; on the other, initially normal ventricles which explode around the 21st–22nd week of gestation resulting in the most severe hydrocephalus. In these cases particularly, close observation of the progress is mandatory to facilitate delivery via cesarian section from the 32nd week on, following a check of the maturity of the lungs.

The second is scheduled delivery, which is of particular importance in gastroschisis or omphalocele, where cesarian section should be carried out as a matter of principle to avoid mechanical lesions of the prolapsed intestinum, to avoid infections, and to make possible an immediate elective operation which should not have the character of emergency. Scheduled delivery following contact with the pediatric surgeon is preferable with all indistinct intra-abdominal abnormalities, such as bowel atresia, abdominal or thoracic cysts, and malformations of the urinary tract and the anorectum, which require immediate postnatal diagnostic procedures.

Table 5. Earliest possible diagnosis and present procedure for malformations which can be diagnosed antenatally

Malformation	Earliest diagnosis in the literature	Procedure/ clarification	Consequences
Anencephalus	9th WP		Interruption of pregnancy
Hydrocephalus (normally)	17th–20th WP	Biweekly checkups	Preterm cesarean (cephalocentesis from 26th WP?, ventriculo-amniotic shunt?)
MMC (normally)	18th/19th WP	Associated malformations Bladder paralysis Kyphosis	Delivery at term Lorber grade IV: interruption of pregnancy before 24th WP?
Cystic renal diseases	13th WP	Oligohydramnios Lung maturity Growth retardation	Normal delivery
Urethral stenosis exit	13th–20th WP	Checkups	Normal delivery (puncture 29th–36th WP?, pyeloamniotic shunt?)
Megalocystis (urethral valves, urethral stenosis)	ca. 13th WP	Pleural hypoplasia	Normal delivery (puncture?, vesico-amniotic shunt?)
Gastroschisis, omphalocele	13th WP	Checkups	Cesarean, if possible at term
Other abdominal malformations	12th WP	Checkups	Normal delivery

MMC, Myelomeningocele; WP, week of pregnancy

With obstructive urinary malformations such as urethral valves with megalocystis, megaureters, and hydronephrosis, which would seem to demand intrauterine treatment, one should wait until more physiological data are available. A preterm delivery from the 32nd week of gestation on seems to be desirable as far as the renal situation is concerned; however, it is dangerous because of lung immaturity. Both risks must be individually considered. Our present procedure in diagnosis of intrauterine malformations is summarized in Table 5.

Discussion

Our investigations show that there has been a considerable increase in the number of antenatal ultrasound examinations, particularly since at least two ultrasound

examinations during pregnancy have been prescribed by law in Bavaria since April 1, 1981. The first is carried out during the 16th–20th week of gestation for disclosure of malformations and the second during the 30th–35th week for control of fetal growth.

Two, however, are certainly not sufficient to observe and guide the course of pregnancy. According to the claims of the *Deutsche Gesellschaft für Ultraschallmedizin* (German Society for Ultrasound Medicine), at least four ultrasound examinations should be performed during pregnancy, and the mother should be transferred to an obstetric center as soon as the slightest suspicion of a malformation arises. The center must be selected according to whether there is a pediatric surgeon available experienced in the correction of congenital malformations. For psychological reasons, it is important to arrange a meeting of the parents, the obstetrician and the pediatric surgeon antenatally in order to relieve fears and answer questions.

Here we must expressly warn against entrusting a senior physician with part-time care of congenital malformations, assuming that the senior physician of a large surgical clinic must automatically have enough experience to handle such cases. To the disadvantage of many malformed infants, it is often kept secret (a) that these senior physicians are frequently unqualified as pediatric surgeons, (b) that they become consulting physicians after 5–6 years, (c) that their successors have to start accumulating their own experience with rare congenital malformations, and finally (d) that care of infants treated this way is divided between the operating general surgeon and the pediatrician doing after care, who is, in turn, not experienced in pediatric surgery. For this reason, a newborn with a definite antenatal diagnosis of a malformation necessitating an emergency operation, such as diaphragmatic hernia, gastroschisis, omphalocele, bowel atresia, urogenital malformations, or myelomeningocele, should be delivered at a time arranged with the pediatric surgeon and immediately referred to a center for pediatric surgery. Unexpected referrals at times when diagnosis and operation can be performed only under difficult conditions should thus be avoided in future.

Our investigations also show that intrauterine operations cannot yet be performed, since despite advances in ultrasound diagnosis, a definite, comprehensive antenatal diagnosis cannot yet be made. Although certain malformations can be safely diagnosed in over 80% of cases by ultrasound, associated malformations impairing prognosis, such as esophageal atresia, Hirschsprung's disease, and congenital heart disease, cannot be excluded. There are still other uncertainties with regard to how far intrauterine findings are treatable and from what point on. Therefore, the results of further animal experiments must be awaited. It is quite probable that intrauterine measures might be useful for extended hydrocephalus, megalocystis with megaureters, and hydronephrosis, for instance. Interesting aspects were shown by Michjeda (Michjeda and Hodgen 1982; Michjeda et al. 1983), who demonstrated that the characteristic paralysis of the lower extremities does not appear in rhesus monkeys with experimentally induced spina bifida if the open vertebral channel is closed with allogenic, agar-stabilized bone tissue in utero. Based on our own investigations of patients with gastroschisis, we are of

the opinion that the urine of the fetus passed into the amniotic fluid is the noxious agent for the bowel slings in gastroschisis as well as for spina bifida. On the one hand, the urine causes edematous swelling of the bowel wall and functional disturbance of ganglionic cells, which, however, recover several weeks postpartum; on the other, it leads to irreversible damage of neuronal structures directly exposed to urine. This means that at least in individual cases of spina bifida neuronal damage is caused not only by inhibitory processes but also secondarily. Early closure of the vertebral arcs could be of therapeutic interest; however, application of this procedure in human beings is at the moment still speculative.

Summary

Based on an increase from 6000 ultrasound examinations done at the First University Gynecological Hospital of Munich in 1969 to more than 13,000 in 1982, and on an increased rate of antenatally diagnosed malformation, the accuracy of antenatal ultrasound diagnosis was investigated. It turned out that correct diagnosis could be made in over 80% of cases. However, this was incomplete in approximately 40% of the cases, since associated malformations of the gastrico-intestinal tract, congenital heart disease, myelomeningocele in correctly diagnosed hydrocephalus, and others were not recognized. False-positive results were found in 17.5% of cases, false-negative ones in 12.7%.

Because of the uncertainty of antenatal diagnosis and ignorance of many physiological parameters, intrauterine surgical treatment cannot be advocated at present. Intrauterine measures are still confined to punctures, administration of drugs, and diagnostic procedures. Further consequences such as termination of pregnancy, psychological aspects, and possible development of intrauterine therapy are discussed.

Résumé

Les auteurs s'interrogent sur la valeur de l'examen échographique anté-natal en se basant sur l'augmentation de ces examens au Premier Hôpital Gynécologique de l'Université de Munich. Ces examens étaient au nombre de 6000 en 1979, et ils sont au nombre de plus de 13000 en 1982. Il a été établi, semble-t-il, qu'un diagnostic correct peut être fait dans plus de 80% des cas, qui était toutefois incomplet dans 40% des cas, du fait de malformations associées du tractus gastro-intestinal, de malformations congénitales du cœur ou de myéloméningocèles dans les hydrocéphalies correctement diagnostiquées, et d'autres malformations qui n'ont pas été diagnostiquées. Les résultats faux positifs étaient de 17,5%, les faux négatifs étaient de 12,7%.

Le traitement chirurgical intra-utérin ne peut être indiqué actuellement à cause de l'insécurité du diagnostic anté-natal et de l'ignorance de nombreux paramètres physiologiques. Les techniques actuelles de traitement intra-utérin sont

limitées à des ponctions, à l'administration de drogues et à des mesures de diagnostic. Des conséquences particulières comme l'interruption de la grossesse, les aspects psychologiques et le développement possible du traitement intra-utérin sont discutés.

Zusammenfassung

Aufgrund der Zunahme der Ultraschalluntersuchungen in der Ultraschallabteilung der Universitätsfrauenklinik München von 6000 im Jahre 1969 auf über 13000 im Jahre 1982 und der dabei gleichzeitig vermehrt pränatal diagnostizierten Fehlbildungen wird die Treffsicherheit der pränatalen Ultraschalldiagnostik überprüft. Dabei zeigt sich, daß in über 80% der Fälle richtige Diagnosen gestellt werden können, daß jedoch bei etwa 40% der Kinder die pränatale Diagnose unvollständig ist, da Begleitfehlbildungen wie Mißbildungen des Magen-Darm-Traktes, Herzfehler, aber auch Myelomeningozelen mit bekanntem Hydrozephalus und anderen Schädigungen nicht erkannt werden. Falsch-positive Befunde kamen in 17,5% der Fälle, falsch-negative bei 12,7% vor.

Aufgrund dieser unsicheren pränatalen Diagnose und der Unkenntnis zahlreicher physiologischer Faktoren kann eine generelle chirurgische intrauterine Therapie zum jetzigen Zeitpunkt nicht befürwortet werden. Intrauterine Eingriffe können sich im Augenblick lediglich auf Punktionen, Verabreichungen von Arzneimitteln und diagnostische Maßnahmen beschränken. Auf weitere Konsequenzen wie Schwangerschaftsunterbrechung, psychische Fragen und mögliche Ansätze einer intrauterinen chirurgischen Therapie wird näher eingegangen.

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Psychological and Ethical Problems Arising for Physicians and Parents from the Prenatal Diagnosis of Malformations

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Undoubtedly, the value of prenatal diagnostic procedures is accompanied by positive psychological effects in the case of a normal fetus. Anxieties are relieved and uncertainties are overcome. A positive psychological effect is also attributed particularly to prenatal ultrasonography by obstetricians and pediatric surgeons even if it leads to the discovery of a malformation in the fetus. The effect is believed to come from several advantages related to early diagnosis. It has been observed that telling the mother and the family may lead to transitory agitation which then can be followed by acceptance of the fact and emotional stabilization. If the physician takes time and care to relate the diagnosis the parents have the opportunity to acquaint themselves with the prospect of having a malformed and perhaps chronically ill child and to prepare themselves for special needs and burdens. In addition, positive psychological effects are expected from reducing the risks of delivery by planned cesarean section, admission to optimally equipped centers, and increased certainty based on calculable risks.

This optimistic stand, however, contrasts with reports given retrospectively by mothers about prenatal diagnoses of fetal anomalies and their disclosure by the physician. Diagnostic unreliability is one complaint which is made not infrequently, referring to the report of an anomaly which either cannot be confirmed at a later examination or is more severe than originally thought. Furthermore, mothers remember the examination as a technical procedure, the diagnosis merely promoting them and their child into an interesting case; they are otherwise left alone with their problem. Taking into account that retrospective reports are most certainly biased by past emotions and later experiences, the fact remains that psychological effects of early prenatal diagnoses have not been studied systematically, and that for the time being the physician has to rely upon anecdotal reports, personal experience, analogies, and his own ability in dealing with difficult situations.

The personal experience of obstetricians and pediatric surgeons in relating the diagnosis of a fetal malformation to the family may be that of destroying the parents' expectation of a healthy child, but explaining the problem and possible interventions as well as the prospect of reducing risks may relieve and compensate for agitation, leading in fact to acceptance of the handicapped child. This would be the optimal development; how frequently it occurs and under what circumstances is not known, but there is reason to doubt that the optimum is the norm.

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Experienced clinicians, at least, seem to approach the situation by anticipating an emotional shock and lasting agitation in mothers and their families and by being prepared to deal with such a possibility. Obstetrical departments have therefore adopted a scheme of appropriate steps for handling eventualities.

1. *Telling the mother.* This should preferably be done by the obstetrician who provides prenatal care during the entire course of pregnancy, and not by the doctor who only makes the technical diagnosis and may not see the mother again. The pediatric surgeon and the pediatrician should be called in to see the mother and the family, give further explanations, and offer help.
2. *Relating the problem in stages.* This does not mean withholding the truth or parts of it. But care should be taken not to tell everything at once, but to make several appointments and to adjust the extent of what is being related to the receptive capacity of the parents.
3. *Presentation of the case at a staff conference.* This does not mean that a "committee" can relieve the individual physician of his or her responsibility, but that discussion within the conference may help to foresee and overcome problems.
4. If serious emotional problems arise, *psychotherapeutic consultation* should be sought.

This scheme offers formal steps to follow, a frame to adhere to, but says nothing about the content and the quality of the procedure. The recommendation usually given in this context to exercise care and empathy remains an empty formula as long as the communication between physician and mother is not such that the emotional situation of the mother can be guided to a specific perspective on the conditions posed by the malformation.

The emotional situation of pregnancy – in contrast to common belief – is not principally or consistently determined by an acceptance of the expected child. Depending upon the personality of the individual woman, pregnancy may solve pre-existing emotional problems or temporarily suppress them. But pregnancy may also activate pre-existing problems or cause new ones. Feelings of ambivalence, anxieties, fears, and unconscious impulses of rejection are normal; i.e., every pregnant woman undergoes a crisis, pregnancy being "a turning point in the life of the individual, leading to acute dysequilibria which under favorable conditions result in specific maturational steps toward new functions" (Bibring 1959).

How easily such imbalance can change to open rejection of the child is demonstrated by the occasional demand for immediate termination of pregnancy in the face of minor anomalies diagnosed by prenatal examination, anomalies which may well be correctable and not even disabling.

Anything that contributes to the maternal personality – life history, psychic structure, and present emotional state – will combine with the mother's social environment (partnership, other family members, occupation) to determine her reaction to pregnancy and to the presence of a malformed child. In the individual case this complex interaction is not predictable, nor can it be channeled into the desirable direction just by a friendly conversation. It is therefore considered

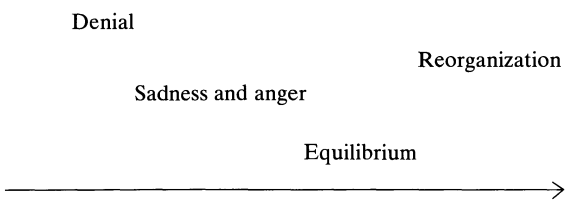

Fig. 1. Model of the sequence of normal parental reactions to the birth of a child with congenital malformations (adapted from Klaus and Kennell 1976)

sensible and necessary, soon after a pregnancy becomes known, to obtain sufficient information about the psychic state and the social situation to identify women at risk for emotional crisis and to try to lessen this risk. Should a complication occur later on, this would then have a protective effect for mother and child.

As far as when parents should be informed of a malformation is concerned, it is most probable that the later they are told, the more negative is their reaction. In the case of a mother and family being told of a malformed child soon after delivery, Klaus and Kennell (1976) and Martinius et al. (1983) found that the confrontation with the malformed child and the discrepancy between the idealized expectation and the reality triggers a sequence of emotional reactions which follows a basic pattern with individual variations (see Fig. 1).

At which emotional stage a parent happens to be is not easy to estimate, because there may be ambivalent feelings and attitudes. Besides, the development in both parents must not necessarily take a synchronous course. Sooner or later, guilt feelings are inevitable. They tend to inhibit further progress, prolonging, for instance, the stage of depression. If, as is usual, guilt feelings are suppressed, this mechanism is liable to produce pathological attitudes toward the child (Martinius et al. 1983).

Of course, physicians themselves react emotionally to being involved in such problematic situations. And not everyone is equally able to accept the challenge and to assume responsibility. To facilitate the engagement of the physician, reference is made once more to the work of Klaus and Kennell (1976), who recommended making use of a number of questions typically asked by mothers and their families, such as: (a) Is the malformation visible?; (b) Is it correctable?; (c) Will there be a need for institutional help?; (d) Is it inherited? With prenatal diagnosis the list would have to be extended by questions relating to prenatal care and delivery. Talking about these purely rational topics nevertheless gives the opportunity to learn more about parental personalities, to perceive and relate feelings, above all to be someone who is involved.

Naturally, one should not expect the exact content of the information given to have been understood. If the recipient has not passed the stage of denial or depression, the information virtually cannot reach him or her. It is a common experience that parents of children born with malformations later do not remember the rational content of the first conversation with their doctor but rather the way in which this conversation was conducted (Richards and McIntosh 1973). In the

early phase, therefore, it is necessary to give the opportunity to ask the same questions repeatedly.

A physician who provides counseling, treatment, and follow-up prenatally is probably well advised if he or she knows what has been reported about prenatal reactions to malformations postnatally. The same applies to other professionals in the field. There is no need for primary psychiatric consultation as long obstetricians, pediatric surgeons, and pediatricians are aware of psychological problems, willing to deal with them, and ready to seek the advice of others if necessary. The initially proposed four-point program should be a useful framework to rely upon.

Aside from psychological problems, prenatal diagnosis raises ethical questions. As always when new diagnostic and therapeutic possibilities are introduced, enthusiasm about the new tool hides its potential drawbacks. Only experience shows that the new method also carries dangers. Prenatal ultrasonography appears to be one of many examples for that sequence of events, namely in three areas: the time limit for legal abortion, the possible overreaction of mothers upon learning of a fetal malformation, and parents being informed of a diagnosis but not of the therapeutic consequences.

The first problem area – the time limit for legal abortion – has been in the public consciousness ever since ultrasonography made it known that as early as 6 weeks gestational age the embryo produces movements which from the 8th week onwards occur systematically generalized and isolated (Ianniruberto and Tajani 1981). In view of these facts, the right of the unborn child to its own life needs to be reconsidered. The present legal status of a fetus has been made obsolete by the new discoveries.

The second problem area – the overreaction, upon being notified of a fetal malformation, in the form of a demand for immediate abortion – is a most embarassing one. Neither can it be ethically justified to leave the parents alone with their decision, nor would it be correct to take the decision entirely from them.

Analogous and equally difficult is the situation in which therapeutic measures cannot be taken, due either to the severity of the malformation or to the fact that none are necessary, at least not prior to and during delivery. In the case of a malformed fetus with little expectation for extrauterine life, the mother and the family must be given adequate opportunity to form their own opinion before termination of pregnancy is presented as imperative. If the mother decides to have the child it would be wrong to accuse her of being irresponsible and thereby exert additional psychological pressure. If an ultrasonographic diagnosis has no necessary therapeutic consequences (atrial septal defect, chondrodystrophy), the question arises to what extent the diagnostic procedure itself (implying the necessity to tell the mother) is ethically sound. The least that can be said is that research should establish soon whether or not harm is being done.

Technical progress does produce advantages. To make full use of them, more than technical know-how is needed. Prenatal diagnosis, particularly with ultrasonography, creates a sensitive psychological situation which can be stabilized if the result is normal and which can be specifically agitated if a malformation is found. Maternal reactions need to be studied.

Meanwhile, making use of prenatal diagnostic techniques, we are obliged to exercise care in recognizing the psychological aspects and in fulfilling the psychological needs originating from such diagnoses.

Summary

Early diagnosis of a malformation has positive effects for the parents. Transitory agitation is followed by acception of the fact and stabilization. Obstetrical departments use a scheme of steps, which starts with talking to the mother and relating the problems in stages and continues with presentation of the case at a staff conference with obstetrician, pediatrician, and pediatric surgeon, and ends with psychotherapeutic consultation if necessary. The emotional situation of pregnancy with a malformed child and confrontation triggering a sequence of emotional reactions including wide individual variations are described.

The use of new prenatal diagnostic techniques requires care in recognizing the psychological aspects and needs originating from unfavorable diagnoses.

Résumé

Le diagnostic précoce d'une malformation a des effets positifs pour les parents. Après une angoisse transitoire, ceux-ci acceptent le fait et une stabilisation survient. Le département d'obstétrique utilise un schéma par étapes qui commence par une discussion avec la mère dans laquelle les différents problèmes sont envisagés dans tous leurs aspects. Ultérieurement, le cas est présenté à une conférence multidisciplinaire comportant un obstétricien, un pédiatre et un chirurgien pédiatrique, et éventuellement, une consultation psychothérapique. Les auteurs évoquent la situation émotionnelle d'une grossesse concernant un enfant malformé et étudient la séquence des réactions psychologiques qui sont particulières à chaque individu.

L'utilisation des nouvelles techniques de diagnostic prénatal requiert une attention particulière dans les aspects psychologiques de l'environnement et nécessitent des précautions spéciales dans les cas de diagnostic défavorable.

Zusammenfassung

Die frühe Diagnose einer Fehlbildung hat eine günstige Wirkung auf die Eltern: einer vorübergehenden Irritation folgt gewöhnlich die Annahme der gegebenen Umstände und eine stabilisierte Einstellung dazu. Geburtshilfliche Abteilungen verwenden ein aus mehreren Schritten bestehendes Schema, das mit dem Gespräch mit der Mutter beginnt, wobei die Problematik in mehrere Einzelaspekte unterteilt wird. Weiterhin folgt die Vorstellung dieses Falls auf einer interdisziplinären Besprechung zwischen Geburtshelfer, Pädiater und Kinderchirurgen sowie

die psychotherapeutische Betreuung (soweit erforderlich). Die emotionale Situation während einer Schwangerschaft mit einem fehlgebildeten Kind und die Konfrontation damit, die eine Reihe von emotionalen Reaktionen von beträchtlicher individueller Variationsbreite auslöst, werden besprochen.

Die Anwendung neuer diagnostischer Methoden erfordert die Erforschung psychologischer Aspekte und der Notwendigkeiten, die aus ungünstigen pränatalen Diagnosen erwachsen können.

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