THE TREATMENT OF OESOPHAGEAL ATRESIA*

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Though both the anatomy and the symptomatology of the first cases of oesophageal atresia were described in detail more than 250 years ago, it was not until 1941 that the first child suffering from this anomaly was successfully operated upon. The mortality rate diminished rapidly during the following years, and, as methods making successful treatment possible were presented, a campaign was started to bring all cases of oesophageal atresia to the surgical wards as soon as possible after birth. That this has not been without effect is shown by the great number of cases now living which make a background to our evaluation of different methods of treatment.

Oesophageal atresia is a fairly common disease. The frequency is said to be 1:2,500-3,000 births. Both prognosis and selection of the method of treatment depend upon the type of malformation dealt with. According to Vogt's (1929) classification there are three principal types, namely, (1) complete absence, (2) atresia without tracheal fistula and (3) atresia with fistula between the trachea and the oesophagus, (a) those with a fistula between the upper segment and the trachea, (b) those with a fistula between the lower segment and the trachea, (c) those with fistulas between both segments and the trachea.

About 90% of all cases have a fistula from the lower oesophageal segment to the trachea and a blind-ending upper segment, that is, the type 3b of Vogt (Fig. 1).

In this connexion I should like to mention a further oesophago-tracheal malformation of which two cases have been recorded in Sweden during the last few years. In 1955, Pettersson, of Gothenburg, published one case where the upper parts of the oesophagus and trachea, including the larynx, were not separated from each other (Fig. 2). The child was operated on and is in good health. I also had a similar case a year ago. In this case, however, the oesophagus and the upper respiratory tract down to the carina were not separated (Fig. 2). An operation was performed through a left-sided cervical incision and a right-sided thoracotomy.

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* The McArthur Lecture delivered at the annual meeting of the British Association of Paediatric Surgeons, 1957.
The patient survived the operation, which lasted eight and a half hours, and was performed with the help of a "narcos" respirator. The child remained in very good condition for a week in the respirator, but then unfortunately died suddenly from some unknown cause.

When the anatomical picture of the malformation is known it is not difficult to understand the clinical picture, and early diagnosis is easy. Characteristically the infant rapidly accumulates mucus in the nose and mouth, and suctioning of the mucus establishes normal breathing. Attempts at feeding are followed by coughing, choking and prompt regurgitation of all fluid given. These symptoms depend on the action of swallowing being blocked by the atresia. A catheter inserted into the oesophagus may demonstrate an obstruction, usually about 10 cm. from the lips. The diagnosis is verified by x-ray investigation after injection of some millilitres of 'lipiodol' through the catheter into the upper oesophagus. The 'lipiodol' should be sucked up after the investigation. Furthermore, the radiographs give valuable information regarding the extent of the pulmonary complications that in these cases usually can be diagnosed as atelectasis or pneumonia. Air in the gastro-intestinal tract indicates a fistula between the lower segment and the trachea. It may be added that it is necessary to remember that oesophageal atresia is often connected with other anomalies, of which congenital heart disease, malformation of the anus or rectum and atresia or stenosis of the small intestine are considered the most common. Sometimes the distribution of the air in the abdomen may suggest atresia of the bowel as an associated, complicating anomaly.

A review of the literature shows that the first important contribution to the surgical treatment of the oesophageal atresia was made by Richter in 1913. He gave a plan for the performance of the surgical procedure. This can be done in two different ways, either by closing the fistula and performing a direct anastomosis between the two oesophageal segments or by a multiple-stage procedure including closing the fistula, putting out the upper segment to a cervical oesphagostomy and making a gastrostomy. Later an oesophagus can be constructed. Richter operated on two children but both died.

Much hard work had to be done until Leven (1941) could report of the life of the first child saved through a multiple-stage procedure. Two years later the first case saved by a direct anastomosis was published by Haight and Towsley (1943). In Europe the first successful operations were reported by ten Kate (1947), Franklin (1948) and Sandblom (1948).

I should like to mention briefly the problems which have arisen during past years since the first successfully treated cases and keep mainly to atresia with fistula from the lower segment.

Since Mixter in 1936 advised the retropleural approach, the transpleural route seems to have been temporarily abandoned as being too dangerous. It became, however, quite natural after the advances in anaesthesia and thoracic surgery to try the transpleural exposure again, with the result that nowadays this method is the one commonly used. By both methods a right-sided approach has proved to be more practical than the one on the left.

The retropleural method means that the mediastinum is entered from a parascapular incision, usually after resection of the second or third down to the sixth ribs.

The transpleural exposure is made by entering the fourth or fifth intercostal space. When the incision into the mediastinal pleura has been made the operation continues in most respects in the same way as with the retropleural method. The azygos vein is divided, the vagus nerve is identified, the lower oesophagus dissected in its upper part and the tracheal fistula is divided and carefully closed. The upper oesophageal pouch is then mobilized and the anastomosis performed.

The disadvantage of the retropleural approach is that it does not easily allow a wide exposure of the mediastinum. The chief advantage, on the other hand, is that should the anastomosis break down the retropleural drainage may save the child and when there is only a partial rupture the anastomosis may heal spontaneously.

The disadvantage of the transpleural approach is that a post-operative insufficiency of the anastomosis may cause a pyopneumothorax which is a very serious complication, but this opening allows a wide exposure of the mediastinum which means that a better anastomosis can be performed. This is considered important as it has made the above mentioned complication rare.

However, several surgeons use the retropleural approach with very good results. Whether the exposure should be retropleural or transpleural is therefore a matter of opinion and still open to debate.

The anastomosis can be done by using one or two layers of interrupted sutures. Many surgeons prefer the Haight-Towsley technique, that is, suturing the whole thickness of the wall of the distal segment to the mucosa of the upper pouch and reinforcing the anastomosis by 'telescoping' the muscular layer of the upper segment around the lower one. With a couple of stitches Swenson (1947) attached the
upper segment to the prevertebral fascia as low down as possible in order to diminish the tension on the anastomosis. Sulamaa, Gripenberg and Ahvenainen (1951) fixed the upper pouch to the lower segment after having closed the oesophago-tracheal fistula without dividing the oesophagus. After that a termino-lateral anastomosis was performed. Ten Kate used a flap from the upper segment for the anastomosis. In order to make sure that the anastomotic line is tight, Sandblom advised injection of saline solution in the upper oesophagus. If a leak could then be observed a closure could be done by one or two sutures more.

Without any doubt several of the various methods described in the literature can be used with success, but in all of them the important thing is to get a tight anastomosis without too much tension. This should be accomplished by a thorough mobilization of not only the upper segment but also as necessary of the lower one.

Often the lumen of the proximal part of the distal segment is very small. Increased diameter can be obtained by dilating it or by making a small longitudinal slit in the wall of the oesophagus. It is, however, well worth noting that even a quite narrow distal segment will usually dilate properly later.

In the first published series a relatively large number of patients had been treated by multiple-stage procedure. This might be explained by the fact that it was considered dangerous to free and mobilize the lower oesophageal segment to a desirable extent, the risk being insufficient nutrition and necrosis of the oesophagus. As it was often found at operation that a gap of several centimetres existed between the ends of the upper and the lower oesophageal segments, it is understandable that the multiple-stage procedure was often resorted to. However, it soon proved that this procedure meant a great deal of trouble and also carried a high mortality. Further, it became clear that the lower segment of the oesophagus could be freed much more than was earlier anticipated. Therefore it gradually became the usual practice to do a direct anastomosis in nearly all cases. It is now considered possible in all cases with oesophago-tracheal fistula. Those cases without fistula have as a rule a very short lower segment and it becomes necessary to pull the stomach up into the chest if a direct anastomosis should be done. However, at the moment there is no agreed method among surgeons. Thus Gross (1953) gave up the multiple-stage procedures in 1945 and prefers drawing the stomach up into the chest when necessary for making it possible to perform a primary anastomosis. Others have, however, tried this method without being able to achieve satisfactory results. Several other methods have been tried in these cases. Thus Sandblom in 1947 tried to construct part of the intrathoracic oesophagus by the transverse colon in a 4-day-old child. The child died. At the last meeting of the British Association of Paediatric Surgeons, Waterston demonstrated excellent results having constructed the intrathoracic part of the oesophagus from a loop of the colon.

Another question is, Should a gastrostomy be performed or not? It has been shown that oral feeding may be started a few days after the primary repair of the oesophagus. It is known that some surgeons keep a small catheter leading from the oesophagus to the stomach after the operation and feed the child through it. It seems, however, that most surgeons still prefer the gastrostomy as the safest procedure. As a matter of fact it can hardly be denied that a gastrostomy gives the best opportunity for the undisturbed healing of an oesophageal anastomosis and I suppose nobody can be seriously criticized if he always used a gastrostomy for feeding the child during the first 10 days post-operatively.

Blood and fluids should be given during operation in accordance with principles generally accepted in order to compensate losses and prevent shock. It has, however, been clarified that the operation represents only part, although an important part, of the treatment, and it might be correct to say that the adequate pre- and post-operative care of the child is the key to a successful outcome.

During the last 10 years many details have been emphasized in the treatment of these children. It is but natural that the early recognition depends on the alertness of those who have to treat the newborn babies. Those persons must also be acquainted with the treatment necessary to save the children from the immediate dangers. It is obvious that it is necessary to aspirate mucus from the pharynx of the child when there is excess and this ought to lead to a suspicion of the diagnosis. At the same time this is the first treatment. After that comes the steps taken in order to avoid complications. Suction should be repeated every quarter to half an hour if needed and feeding by mouth should be stopped. The child ought to be placed in a half-prone position in order to avoid regurgitation and aspiration of gastric contents.

If the preliminary diagnosis has been made shortly after birth and the child is in good condition there is no need for administration of parenteral fluids before the child is taken to a surgical department. In all cases it seems wise to start treatment
with antibiotics and to give vitamin K prophylactically.

When the patient is being transported, the person accompanying the child must be in a position to provide suction.

When the patient has been admitted to the surgical department and diagnostic measures are completed, treatment has to be decided upon. It must then be clear that though the case itself may be described as an emergency, it does not mean that the operation should be carried out immediately. On the contrary, under a diligently-performed prophylaxis against aspiration, a child who on admission is seriously ill can be brought into good condition and be given a fair chance to withstand the surgical procedure.

One way to establish this pre-operative care is to place the child in an incubator with the temperature controlled in saturated humidity. The pharynx is sucked out every quarter of an hour if needed. Oxygen is given when indicated. Treatment with antibiotics is continued or instituted if not given earlier. When indicated the child is given fluids parenterally, keeping in mind the warning against overhydration. We have followed Gross's advice to give 10% glucose in water intravenously to combat any existing ketosis. The amount should not exceed 20 ml per kg. body weight every 12 hours. Generally this pre-operative treatment for a few hours will be enough; it may be given for 12 to 24 hours or even longer when the infant is in bad condition.

After the operation the child is brought back to the incubator which is properly warmed and humidified. There the child lies quite naked and it is possible to make continuous observations regarding its way of moving and breathing and its colour. Often the pharynx has to be sucked out frequently during the first few days. After that the child apparently is often able to swallow most of its saliva. Treatment with antibiotics is continued until the pulmonary complications are under control. In former years the administration of oxygen during several days post-operatively had been advised. Today we prefer to use only air in the incubator routinely. Humidity seems to be of the greatest value in the treatment and prophylaxis of pulmonary complications. The child must be kept from being overhydrated, and the parenteral fluid regulated, starting with about 60 ml per kg. body weight per day and increasing gradually under weight controls. Gastrostomy feeds are started about 24 hours after operation and are as a routine continued for eight to 10 days. An x-ray investigation is then done and if it shows a good passage without fistula oral feeds are initiated. The gastrostomy tube is kept in for six to eight weeks. If there is no stenosis demonstrable the tube is then removed. After that the gastrostomy will usually close spontaneously.

In spite of the fact that the frequency of complications is diminishing due to safer methods of operation and better understanding of the pre- and post-operative care, we must always be prepared to face complications from the respiratory tract. Repeated radiographs are often necessary to determine the therapeutic effect on atelectasis and pneumonias. Repeated suction of the trachea and the bronchi may be necessary to eliminate a stagnating secretion which might otherwise rapidly kill the infant. Thus team work between the nurses, surgeons, anaesthetists and radiologists is necessary for the achievement of a good result.

In a certain number of cases stenosis of the anastomotic line develops, sometimes early in the post-operative course and sometimes later. This complication is usually not very dangerous if observed and treated by dilatation.

The most feared complication is of course rupture of the anastomosis. Sometimes it ruptures partially and may gradually develop into a pyopneumothorax, sometimes, however, the child deteriorates quickly.

I am unable to discuss the merits of different methods of anaesthesia in oesophageal atresia. Our anaesthetists have, in several cases, avoiding intratracheal tubes, administered cyclopropane-oxygen mixture through a tightly fitting face mask. We have thought this procedure less irritating for the larynx, but I know that there is no agreement about the advantages of this method.

Regarding the unusual types of oesophageal atresia, we have treated a few cases with rather disappointing results. However, at an analysis of these cases I have found nothing worth adding to that mentioned earlier.

In order to give an idea of the effect of our treatment I should like to show our results during the last five years (Table 1). The series consists of all cases of oesophageal atresia treated during that

<table>
<thead>
<tr>
<th>TABLE 1</th>
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<tbody>
<tr>
<td>105 CASES OF OESOPHAGEAL ATRESIA</td>
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<tr>
<td>----------------------------------</td>
</tr>
<tr>
<td>1951-1956</td>
</tr>
<tr>
<td>----------</td>
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<tr>
<td>Atresia with fistula from the lower oesophageal segment</td>
</tr>
<tr>
<td>Other types of atresia</td>
</tr>
<tr>
<td>Total</td>
</tr>
<tr>
<td>11</td>
</tr>
<tr>
<td>105</td>
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time at the Carolina Hospital, Stockholm, the Crown Princess Louisa’s Hospital, Stockholm and at the University Hospital, Lund.*

From Table 1 it is apparent that of 105 cases 101 have been operated upon with 46 deaths. That is an overall mortality of 44%. Ninety-four cases belonged to the group of atresia with fistula from the lower segment with a mortality of 41%. Eleven cases having some other type of the malformation were treated with a high mortality rate. The poor results in this small group show that we have not found a satisfactory method for these cases.

When discussing the results and the possibilities of getting better results, it has to be borne in mind that this series consists of many unfavourable cases. Among the factors of importance for the prognosis there are three uncontrollable factors. The first one is the type of anomaly, the second one is the degree of immaturity and the third is the presence or absence of associated anomalies. It is well known that these factors greatly affect the prognosis. Thus it will seem adequate when estimating the results to divide the series into the following groups shown in Table 2.

It is not astonishing to find a high mortality rate among the immature infants weighing less than 2,500 g., nor among those with serious associated anomalies. However, some of these anomalies can be corrected and others may be of minor importance for the immediate result. Therefore a certain number of these cases survive. Table 2 shows that among the cases of atresia with fistula between the lower segment and the trachea those with a weight of more than 2,500 g. and with no associated anomalies have a survival rate of 82% (the mortality rate 18%). This is gratifying as all those children can be expected to develop into functionally normal individuals.

### Table 2

<table>
<thead>
<tr>
<th>Weight</th>
<th>Number</th>
<th>Not Operated</th>
<th>Deaths</th>
<th>Mortality (%)</th>
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<tbody>
<tr>
<td>&gt; 2,500 g.</td>
<td>44</td>
<td>1</td>
<td>8</td>
<td>18</td>
</tr>
<tr>
<td>&lt; 2,500 g.</td>
<td>20</td>
<td>2</td>
<td>11</td>
<td>58</td>
</tr>
<tr>
<td>&gt; 2,500 g.</td>
<td>19</td>
<td>2</td>
<td>11</td>
<td>65</td>
</tr>
<tr>
<td>&lt; 2,500 g.</td>
<td>11</td>
<td>1</td>
<td>7</td>
<td>70</td>
</tr>
<tr>
<td>Total</td>
<td>94</td>
<td>4</td>
<td>37</td>
<td>41</td>
</tr>
</tbody>
</table>

* The series from the Carolina Hospital (62 cases) has kindly been put at my disposal by Th. Ehrenpreis and N. O. Ericsson, and the cases (23) from the Crown Princess Louisa’s Hospital by G. Ekström.

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**References**


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