Esophageal atresia without distal tracheoesophageal fistula: high incidence of proximal fistula

Klaas(N) M.A. Bax*, Anne Margot C. Roskott, David C. van der Zee

Department of Pediatric Surgery, Wilhelmina Children’s Hospital, University Medical Center Utrecht, 3508 AB Utrecht, The Netherlands

Key words:
Esophageal atresia;
Tracheoesophageal fistula;
Incidence;
Imaging

Abstract

Background: This retrospective study was performed to test our suspicion that the incidence of esophageal atresia with proximal fistula in our institution is much higher than is generally reported.

Methods: The charts of all patients with esophageal atresia and/or tracheoesophageal fistula admitted in the period 1982 to 2000 were analyzed. The type of atresia and/or tracheoesophageal fistula was noted, and the relative incidence was calculated and compared with the relative incidence in a cumulative series of 3492 patients taken from 9 published studies.

Results: In the period under study, 123 patients with esophageal atresia and/or tracheoesophageal fistula were identified. The relative incidence of esophageal atresia without distal fistula was statistically not different (10.6% in the present series against 8.49% in the reference group). A statistically significant difference in the relative incidence of esophageal atresia with proximal fistula, however, was found: 5.69% in the present series against 1.05% in the reference group (P < .0001). Looking at the subgroup of patients without a distal fistula, more than half of the patients did have a proximal fistula.

Conclusions: The relative incidence of esophageal atresia with proximal fistula in this series of children with esophageal atresia and/or tracheoesophageal fistula is significantly higher than reported in the literature. This is on the account of the subgroup of patients without a distal fistula in which the incidence of a proximal fistula is more than 50%. Especially in this subgroup, the existence of a proximal fistula should be ruled out preoperatively.

© 2008 Elsevier Inc. All rights reserved.

The reported incidence of esophageal atresia without distal but with proximal tracheoesophageal fistula varies between 0.4% and 3% (Table 1) [1-9]. As we had the impression that the incidence at our institution is consider-ably higher, we decided to study the relative incidence of the various types of tracheoesophageal fistula retrospectively and to compare these with the literature.

1. Patients and methods

The charts of all children with esophageal atresia and any type of congenital tracheoesophageal fistula treated at our institution in the period 1982 to 2000 were studied. The type
### Table 1  Relative incidence of various types of esophageal atresia and/or tracheoesophageal fistula

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Type A</td>
<td>n 37.00</td>
<td>25.00</td>
<td>29.00</td>
<td>82.00</td>
<td>39.00</td>
<td>8.00</td>
<td>26.00</td>
<td>6.00</td>
<td>8.00</td>
<td>260.00</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Type B</td>
<td>Proximal fistula 11.00</td>
<td>2.00</td>
<td>2.00</td>
<td>9.00</td>
<td>2.00</td>
<td>1.00</td>
<td>4.00</td>
<td>2.00</td>
<td>4.00</td>
<td>37.00</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Type C</td>
<td>Distal fistula 488.00</td>
<td>329.00</td>
<td>178.00</td>
<td>916.00</td>
<td>441.00</td>
<td>146.00</td>
<td>317.00</td>
<td>86.00</td>
<td>113.00</td>
<td>3014.00</td>
<td>101</td>
<td></td>
</tr>
<tr>
<td>Type D</td>
<td>Proximal and distal fistula 1.00</td>
<td>2.00</td>
<td>5.00</td>
<td>7.00</td>
<td>5.00</td>
<td>0.00</td>
<td>10.00</td>
<td>6.00</td>
<td>3.00</td>
<td>39.00</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Type E</td>
<td>H fistula 32.00</td>
<td>569.00</td>
<td>13.00</td>
<td>13.00</td>
<td>44.00</td>
<td>12.00</td>
<td>4.00</td>
<td>15.00</td>
<td>4.00</td>
<td>5.00</td>
<td>142.00</td>
<td>8</td>
</tr>
<tr>
<td>Type Sum</td>
<td>% 99.90</td>
<td>99.98</td>
<td>523</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type A</td>
<td>Pure esophageal atresia 6.5</td>
<td>6.7</td>
<td>13</td>
<td>7.7</td>
<td>7.8</td>
<td>5.00</td>
<td>7</td>
<td>6</td>
<td>6</td>
<td>7.44</td>
<td>4.87</td>
<td></td>
</tr>
<tr>
<td>Type B</td>
<td>Proximal fistula 1.9</td>
<td>0.5</td>
<td>1</td>
<td>0.8</td>
<td>0.4</td>
<td>0.6</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>1.05</td>
<td>5.69</td>
<td></td>
</tr>
<tr>
<td>Type C</td>
<td>Distal fistula 85.7</td>
<td>88.7</td>
<td>78</td>
<td>86.6</td>
<td>88.2</td>
<td>91.8</td>
<td>85.2</td>
<td>86</td>
<td>85</td>
<td>86.31</td>
<td>82.11</td>
<td></td>
</tr>
<tr>
<td>Type D</td>
<td>Proximal and distal fistula 0.1</td>
<td>0.5</td>
<td>2</td>
<td>0.6</td>
<td>1</td>
<td>0</td>
<td>2.6</td>
<td>6</td>
<td>2.3</td>
<td>1.10</td>
<td>0.81</td>
<td></td>
</tr>
<tr>
<td>Type E</td>
<td>H fistula 6</td>
<td>3.5</td>
<td>6</td>
<td>4.2</td>
<td>2.4</td>
<td>2.5</td>
<td>4</td>
<td>4</td>
<td>3.7</td>
<td>4.00</td>
<td>6.5</td>
<td></td>
</tr>
</tbody>
</table>
of atresia and/or fistula was concluded from imaging studies, operative reports, and follow-up. The final diagnosis of a proximal fistula was made by tracheoscopy in all patients. The relative incidence of the various types was compared with a composite reference series of esophageal atresia and/or tracheoesophageal fistula. Series of more than 100 cases, stating precisely the relative incidence of the various forms of esophageal atresia and/or tracheoesophageal fistula, were selected from the literature for the reference series.

The information was entered in Statistical Products and Service Solutions 12.0.1 for Windows (SPSS, Chicago, IL). For comparison, a binomial exact test was used.

2. Results

One hundred three children with esophageal atresia and/or tracheoesophageal fistula were treated at our institution during the period under study. Thirteen children (10.4%) had an esophageal atresia without distal tracheoesophageal fistula. From these 13 children, 7 (5.6% of the total number) had esophageal atresia with proximal fistula and 6 children (4.8%) had pure esophageal atresia.

The reference series consists of 9 separate series and comprises 3492 patients (Table 1). The incidence of esophageal atresia without distal fistula in the reference series was 8.5%. This number is made up of 1.05% esophageal atresia with proximal fistula and 7.44% pure esophageal atresia.

The relative incidence of esophageal atresia with proximal fistula in the present series is therefore much higher ($P = .0001$) than in the literature. Looking at esophageal atresia without distal fistula, the incidence of having a proximal fistula is more than 50% in the present series against 12.4% in the cumulative series.

3. Discussion

Although the relative incidence of esophageal atresia without distal fistula in the present series does not differ significantly from the reported incidence in the literature (10.4% vs 8.6%), the relative incidence of esophageal atresia without distal fistula but with proximal fistula in the present series is significantly higher (5.69% vs 1.05%) [1-9]. This finding suggests that the lower incidence of esophageal atresia with proximal fistula in the literature is the result of missed diagnosis. When an esophageal atresia without distal fistula is repaired, extensive dissection of the proximal pouch is customary. During this extensive dissection, the proximal fistula may be severed unnoticed, more so because the proximal fistula is never located at the end of the proximal pouch. Whenever the surgeon during the dissection of the upper pouch feels that at some point the esophagus and trachea share a common wall or whenever the trachea is entered during the dissection, a proximal fistula should be suspected.

Although the relative incidence of a proximal in combination with a distal fistula is around 1% (1% in the present series and 1.1% in the cumulative series), one could argue about routine diagnostic tracheoscopy in esophageal atresia with distal fistula. Two reports, however, have been published with much higher relative incidence of esophageal atresia with double fistula. Dudgeon et al reported an incidence of 5.3% in a series of 170 patients and Johnson et al an incidence of 7.7% in a series of 78 patients [10,11]. It seems therefore wise to exclude a proximal fistula in all patients with esophageal atresia.

When the subgroups of pure esophageal atresia and esophageal atresia with proximal fistula are taken together, then the relative incidence of a proximal fistula in the literature is about 12.5% (6.4% to 33.3%). In the present series, 53% of the patients without distal fistula had a proximal fistula. This underscores the need for excluding a proximal fistula in children with esophageal atresia without distal fistula, more so because it is customary to delay esophageal repair for 2 to 3 months. Repeated aspiration of saliva through the undiagnosed proximal fistula may result in pulmonary problems. If a proximal fistula is diagnosed in these patients and respiratory problems do occur, the time schedule of the esophageal repair should be brought forward.

As a general rule, the presence of a proximal fistula should, nowadays, be ruled out preoperatively and not perioperatively. Preoperative diagnosis can be done by means of a contrast study of the upper esophagus or by means of tracheoscopy, but even then the diagnosis of a proximal fistula may be missed [12,13]. Recently, ultrasound evaluation of esophageal atresia and tracheoesophageal fistula has been advocated for delineation of the exact pathology [14]. The disadvantage of such an examination remains that its value is operator dependent. Computed tomography and magnetic resonance scanning have entered the diagnostic arena in esophageal atresia [15]. The ionizing radiation in computed tomography scanning and the need for general anesthesia in magnetic resonance scanning are drawbacks. Nevertheless, the time has come to make a detailed preoperative picture of the whole situation before planning surgery: the presence and location of fistulae, the untouched distance between the esophageal ends, aortic arch anomalies, and tracheal anomalies such as stenosis and tracheomalacia. All these findings may influence treatment, for example, approach through the opposite chest when the aorta arch rotates downward on the right. Extensive mobilization of the proximal esophagus should only be carried out when absolutely needed, for example, when a proximal fistula has been diagnosed preoperatively. It has been shown that the recurrent laryngeal nerves gives motor branches to the upper esophageal pouch in esophageal atresia [16]. These branches will undoubtedly be damaged during such a mobilization, which may contribute to esophageal dysmotility. Swallowing disorders have been diagnosed on
videofluoroscopy and videomanometry in patients with repaired esophageal atresia \[17,18\]. The extensive transcervical mobilization of the esophagus in case of an H fistula may also well be responsible for the high incidence of motility problems seen in these patients \[19\].

**References**


