Pediatr Surg Int (1990) 5: 103–105

Pediatric Surgery
International

Esophageal atresia and right aortic arch

Right or left thoracotomy?

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Accepted September 5, 1989

Abstract. A right aortic arch occurs in 5% of patients with esophageal atresia. Its presence has significant implications; there is a high association of other anomalies, especially cardiac types. Repair of the atresia has been regarded as difficult with the usual approach through the right chest. We report our experience with five cases of right aortic arch and esophageal atresia treated in the past 5 years. In four cases this association was not recognized preoperatively and the right aortic arch was not an obstacle in repairing the atresia through the standard right thoracotomy. In one case the right aortic arch was diagnosed before surgery; repair was done through the left chest.

Key words: Esophageal atresia – Tracheoesophageal fistula – Right aortic arch

Introduction

A right (Rt) aortic arch occurs in about 5% of infants with esophageal atresia and tracheoesophageal fistula (TEF) [2, 3, 5, 9, 13]. These patients have a high incidence of associated cardiac anomalies and occasionally vascular rings [2, 6, 9, 13]. Repair of esophageal atresia has been regarded as difficult with the usual approach through the right chest [1, 5]. We report our recent experience with five cases of esophageal atresia and distal TEF associated with a Rt aortic arch, four of which were first recognized at Rt thoracotomy (Table 1).

Patients and methods

Case 1. A 900-g female newborn with esophageal atresia and distal TEF was found to have a Rt aortic arch during a Rt extrapleural thoracotomy performed at 1 day of age. The lower esophageal fistula connected to the left (Lt) mainstem bronchus. Uneventful division of the fistula and end-to-end esophageal anastomosis was done. A postoperative cardiology evaluation revealed no other cardiac anomalies. She recovered well and had no postoperative complications. After 2 years she was lost to follow-up.

Case 2. A 2200-g female newborn with esophageal atresia and distal TEF was suspected of having a Rt aortic arch by plain chest X-Ray (Fig. 1). The Rt aortic arch was confirmed by fluoroscopy and CT scan of the chest (Fig. 2). Division of the fistula and end-to-end esophageal anastomosis was done through a Lt extrapleural thoracotomy. She had an uncomplicated recovery and is well 2 years following surgery. Moderate gastroesophageal reflux has responded to medical management. An associated ventricular septal defect (VSP) required no treatment.

Case 3. A 2800-g newborn female had perinatal asphyxia and required immediate resuscitation at birth with endotracheal intubation. During resuscitation, a posterior pharyngoesophageal tear was produced. She had esophageal atresia and distal TEF associated with multiple anomalies including a complex cardiac anomaly, choanal atresia, and coloboma.

A Rt extrapleural thoracotomy was done; a Rt aortic arch was evident. A nasogastric tube previously passed was found to be free in the mediastinum through a high posterior esophageal perforation. The distal esophagus lay to the Lt of the aorta with the fistula ending in the Lt main stem bronchus. The fistula was divided and closed. Because of the pharyngoesophageal perforation only minimal mobilization of the upper pouch was done. The upper pouch was not opened and anastomosis was not attempted; the esophageal ends were brought together with two 4–0 nylon sutures, hoping for spontaneous fistulization. The area was appropriately drained. A Stamm gastrostomy was done. A tracheostomy was performed because of bony choanal atresia. Spontaneous fistulization of the two esophageal ends was noted after 2 months; endoscopic stringing with subsequent dilatations resulted in a satisfactory esophageal lumen. Repair of the choanal atresia was done at the same time. At 3 months of age a Nissen fundoplication was necessary to treat severe gastroesophageal reflux.

Despite the fact of the surgical observation of a Rt aortic arch, this anomaly was not totally demonstrated radiologically. Angiography done for investigation of the cardiac anomalies demonstrated the aortic arch to be more to the middle than to the Rt, with the aorta descending towards the Rt side.

This infant died at 6 months of age from progressive cardiorespiratory failure. An autopsy was not done.

Case 4. A 2080-g newborn male underwent repair of esophageal atresia and distal TEF through a Rt extrapleural thoracotomy. A Rt aortic arch was found during surgery. Division of the fistula and end-to-end anastomosis was easily accomplished. Postoperative recovery was uneventful. An esophagram done at 1 year of age was normal. Cardiologic evaluation revealed no associated anomalies.

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Table 1. Summary of cases and results

Case no.	Birth weight	Sex	Anomaly	Associated anomalies	Approach	Result
1	900 g	F	Esophageal atresia and distal TEF	VSD Rt aortic arch	Rt chest	good
2	2200 g	F	Esophageal atresia and distal TEF	VSD Rt aortic arch	Lt chest	good
3	2800 g	F	Esophageal atresia and distal TEF	Microcephaly, complex cardiac anomaly and VSD, choanal atresia, coloboma, Rt aortic arch	Rt chest	died at 6 months of age from associated anomalies
4	2080 g	M	Esophageal atresia and distal TEF	Rt aortic arch	Rt chest	good
5	2030 g	M	Esophageal atresia and distal TEF	VATER Syndrome VSD Rt aortic arch	Rt chest	good

Case 5. A 2030-g newborn male was found to have VATER syndrome [10] (vertebral anomalies, anorectal agenesis, esophageal atresia and distal TEF, and radial dysplasia). Because of respiratory problems a gastrostomy only was done on day 1; 2 days later a Rt extrapleural thoracotomy was done and a Rt aortic arch was found. The distal TEF was divided and ligated but because of a long gap between the esophageal segments it was elected not to mobilize the upper pouch at this stage. An anoplasty for a low imperforate anus was done at the same time.

At 3 months of age, an uneventful end-to-end esophageal anastomosis was accomplished through a Rt transpleural thoracotomy; an upper circular esophageal myotomy [7] was necessary in order to bridge the long esophageal gap.

At 2 years of age a Nissen fundoplication was done for severe gastroesophageal reflux. An asymptomatic VSD required no treatment.

At 5 years of age the boy is well but has esophageal dysmotility and occasional pulmonary problems due to aspiration.

Discussion

The presence of a Rt aortic arch in association with esophageal atresia has significant implications. There is a high incidence of associated anomalies, especially cardiac malformations [2, 6, 9, 13]. It can influence the surgical approach, since the repair has been regarded as easier when done through the chest opposite the aortic arch [5]. If an unsuspected Rt aortic arch is found during repair of esophageal atresia through the Rt chest, the surgeon should recognize this anomaly to avoid complications. Despite its significant implications, this association is not mentioned in the most recent edition of a pediatric surgery textbook [11].

In one of our five cases the preoperative diagnosis of Rt aortic arch was made; repair of the esophageal atresia was done through the Lt chest. In the other four the Rt

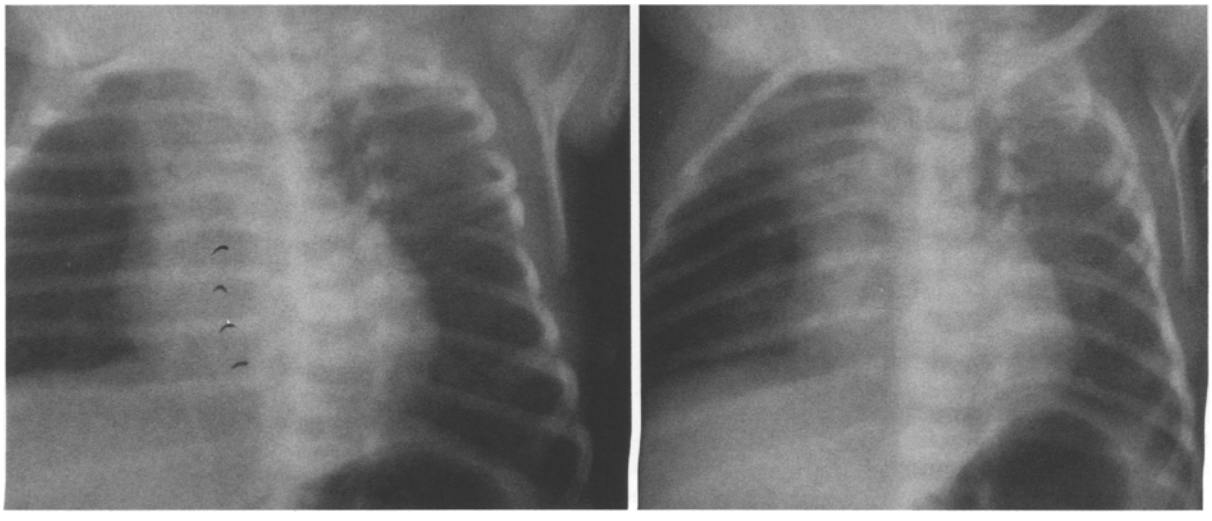

Fig. 1. Chest X-ray film demonstrating the right-sided aortic arch (outlined by black marks)

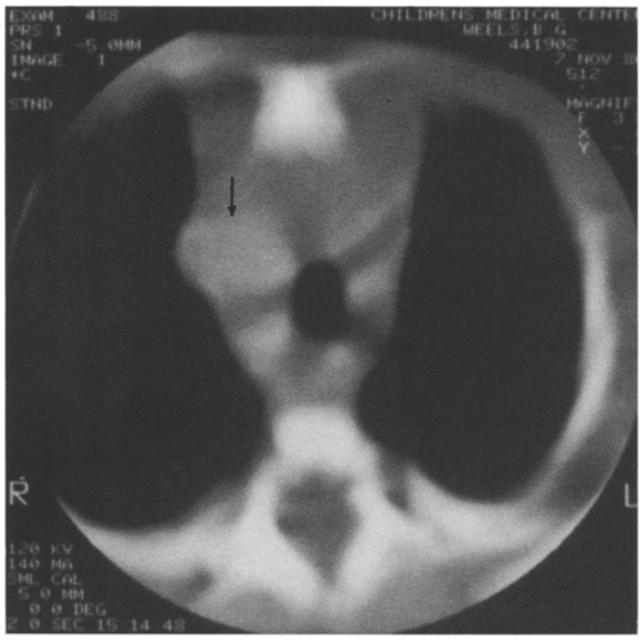

Fig. 2. CT scan of the chest showing the right-sided aortic arch (*arrow*)

aortic arch was recognized during surgery through a Rt thoracotomy. In two of those cases the esophageal repair was easily accomplished. In one infant with a pharyngo-esophageal perforation the tracheoesophageal fistula was divided and closed. Exposure of the upper pouch was easy, but because of the perforation with contamination we elected not to open the upper pouch, instead performing suture approximation of the two ends as described by Shafer and David [12] and Marshall [8]. Fistulization between the two ends occurred and the esophagus was subsequently dilated to a satisfactory lumen.

In one infant with VATER syndrome [10] only a gastrostomy was done initially because of respiratory complications; 2 days later, through a Rt thoracotomy, a Rt aortic arch was discovered. The TEF was easily divided but the anastomosis was not attempted because of a long gap between the esophageal segments. At 3 months of age and knowing that the infant had a Rt aortic arch, the esophageal atresia was approached and repaired through a Rt transpleural thoracotomy; a Livaditis circular myotomy [7] was needed to bridge the long gap. The decision for a right-sided approach was made because during the previous division of the fistula it had been observed that the lower esophageal end was positioned toward the Rt chest. In this case the difficulty in doing the anastomosis was related to the long esophageal gap and not to the Rt aortic arch.

Based on our experience, we conclude that if a Rt aortic arch is suspected preoperatively, the diagnosis can be confirmed by plain X-ray films [1, 9] or a CT scan; angiography is more invasive and unnecessary. Other diagnostic methods such as preoperative endoscopy, magnetic resonance imaging, and ultrasound may be useful, but these procedures were not done in our patients. When the presence of a Rt aortic arch is confirmed, repair is probably easier through the opposite chest [5]. If an unsuspected Rt aortic arch is found during repair of esophageal atresia through a Rt thoracotomy, division of the TEF and esophageal anastomosis can be accomplished and should be attempted unless there are other contraindications in order to avoid an unnecessary second thoracotomy through the opposite chest. If there is a long gap or other anatomical or technical problem, it should be treated appropriately, regardless of whether there is a Rt or Lt aortic arch. If a second thoracotomy is needed, it can be done again through the Rt chest as demonstrated in one of our cases. It is interesting that the first successful repair reported by Cameron Haight was done through the Lt chest in an infant with a Lt aortic arch [4].

Mobilizing the aortic arch is unnecessary and can be dangerous [5]. The upper pouch should be mobilized outside the arch to avoid a potential vascular ring. When dissecting the TEF, care most be taken to avoid confusing the aorta with the esophagus; the aorta in these cases is usually to the Rt of the esophagus but will enter the abdomen through a normally positioned aortic hiatus in the diaphragm [6].

We believe that the presence of a Rt aortic arch is not a contraindication to attempt repair of esophageal atresia through the Rt chest. The decision should be based on the surgeon's experience and personal preference.

Infants with Rt aortic arch should have a complete cardiologic evaluation.

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