Repair of oesophageal atresia with tracheo-oesophageal fistula associated with dextrocardia through right-sided thoracotomy approach

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Right thoracotomy for oesophageal atresia (OA) with dextrocardia is technically challenging due to the heart being in the operative field, and also due to the possibility of right-sided aortic arch. We report a neonate with long-gap OA with tracheo-oesophageal fistula (TOF), dextrocardia, and left-sided aortic arch who was successfully operated by using right thoracotomy. On the basis of our review of the literature and our experience from this case, we found that conventional right thoracotomy is appropriate for OA+ TOF associated with dextrocardia and left aortic arch, and left thoracotomy in dextrocardia and right aortic arch. *Ann Pediatr Surg* 12:109–110 © 2016 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2016, **12**:109–110

Keywords: dextrocardia, oesophageal atresia, right thoracotomy, tracheo esophageal fistula

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Received 15 February 2016 **accepted** 9 April 2016

Background

The conventional surgical approach for oesophageal atresia (OA) with tracheo-oesophageal fistula (TOF) repair is by using right thoracotomy. Associated dextrocardia or right-sided aortic arch (RAA) makes the procedure technically challenging. There has been considerable debate about the optimal surgical approach for OA with associated RAA. However, recent studies reported successful repair by using the conventional right thoracotomy approach [1,2]. Dextrocardia is extremely rare in OA with an incident of around 1% [3]. Unlike RAA, where echocardiography is unreliable for preoperative diagnosis [1,2], dextrocardia is easily diagnosed on using echocardiography as well as a chest radiograph. As the association between OA + TOF and dextrocardia is rarely encountered, it is not extensively studied. Currently, there are only few case reports in the literature [4].

Case presentation

A male neonate born preterm at 32 weeks of gestation and weighing 1.7 kg was postnatally diagnosed to have OA + TOF, dextrocardia, vertebral defect and 13 pair of ribs (Fig. 1).

Echocardiogram confirmed dextrocardia and revealed a structurally normal heart with a left aortic arch (LAA). An ultrasound scan of the abdomen revealed bilaterally normal kidneys with situs solitus.

A right-sided thoracotomy was carried out. Good exposure of distal oesophagus was achieved after retracting the heart. The surgery was intermittently interrupted by haemodynamic instability caused by impedance to venous return because of cardiac retraction. However, all such episodes resolved spontaneously when the retractors were removed.

The fistula was identified between the distal oesophagus and the right main bronchus and subsequently ligated and divided. The gap between the upper and the lower pouch was four vertebral bodies. The attempt to perform primary anastomosis was unsuccessful. Therefore, the divided fistula was tucked to the thoracic vertebrae with a prolene stitch and a gastrostomy was carried out for feeding and a Replogle tube was kept *in situ*.

Gap assessment was carried out at 14 weeks of age when the baby weighed 3.8 kg. A bougie was introduced to the upper pouch and a flexible oesophagoscope through gastrostomy to the lower pouch under an image intensifier. This was followed by an uneventful delayed primary repair of the OA through a right-sided thoracotomy using the same old scar.

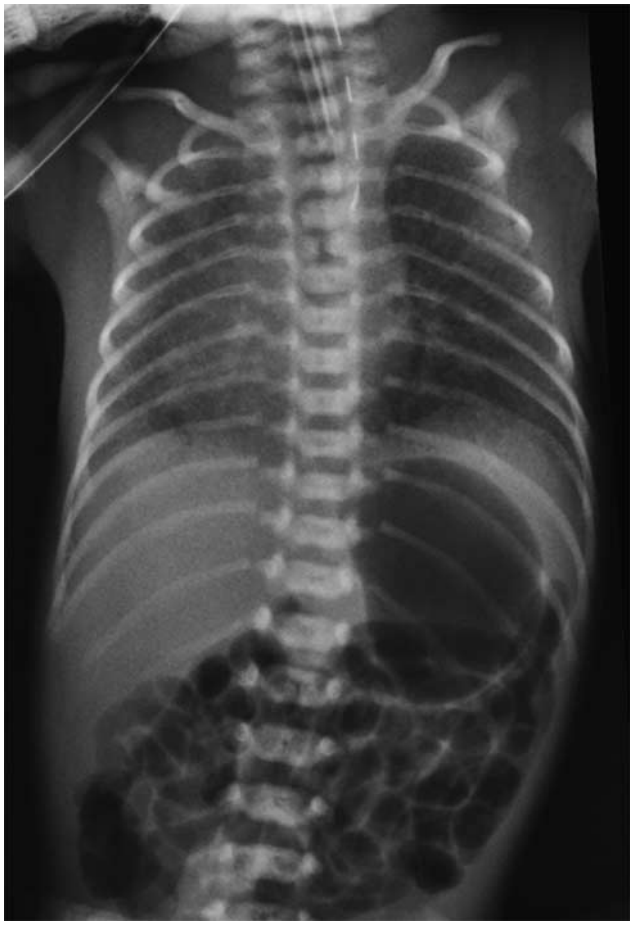
Postoperatively, the child developed anastomotic stricture that required regular dilatations. He also had fundoplication for reflux.

Discussion

The first successful extra-pleural primary repair of OA + TOF was performed by using left thoracotomy in 1941. However, the preferred approach soon changed to right thoracotomy because of better visibility of the lower pouch [5]. While approaching the oesophagus from the left, the visibility of the proximal oesophagus is similar to that of right thoracotomy. However, the distal oesophagus is in a narrow groove between the pericardium and descending aorta making it difficult to identify [6]. In addition, in the left-sided approach, the presence of the aorta prevents the exposure of the lower segment until the aorta has been mobilized by ligation and division of the upper intercostal arteries [5]. In the right-sided approach, the azygos vein can be easily divided to ease fistula ligation without any serious consequences. Therefore, in-situ inversus totalis, due to the presence of the heart and aortic arch on the right, left thoracotomy appears to be the less challenging option. In previously reported cases of OA + TOF with associated situs inversus totalis, left thoracotomy was preferably used [4].

1687-4137 © 2016 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000482858.77300.4d

Fig. 1



The figure demonstrates intubated baby with Replogle tube *in situ*, dextrocardia, vertebral anomaly on fifth thoracic vertebra, and 13 pairs of ribs. Normal position of the liver and stomach supports situs solitus.

However, in our case, dextrocardia was not a part of situs inversus totalis and the aortic arch was on the left side. In neonates with dextrocardia with LAA, the surgical approach is controversial. The choice is between the familiarity with the conventional approach and the anatomical advantage. In OA + TOF with RAA, although there is anatomical disadvantage, some surgeons still prefer conventional right thoracotomy because of familiarity with this approach [1,2]. Therefore, if LAA is diagnosed preoperatively, OA + TOF could be successfully repaired through conventional right thoracotomy even in the presence of dextrocardia, as demonstrated in our case.

Preoperative echocardiogram is unreliable in diagnosing the position of the aortic arch making preoperative decision-making difficult [2]. There are three basic cardiac malpositions: (1) situs inversus totalis, (2) situs solitus with dextrocardia and (3) situs inversus with levocardia. In all the malpositions, the position of the aortic arch corresponded more with the position of the abdominal viscera than with the cardiac position [7]. For example, neonates with situs inversus and levocardia are more likely to have a RAA despite of the normal cardiac position. Therefore, we suggest that an abdominal ultrasound should be used as an adjunct to echocardiogram to determine the position of the aortic arch.

Our case had 13 pairs of ribs, which is associated with long-gap OA + TOF [8]. Although we were initially unable to repair the atresia, primary repair is still possible and should be attempted.

On the basis of our review of the limited available literature and our experience from this case, we found that conventional right thoracotomy is appropriate for OA + TOF associated with dextrocardia and LAA. Paediatric surgeons are more familiar with this approach and the surgery is feasible without any major technical difficulties. However, preoperative confirmation of LAA with echocardiogram and abdominal ultrasound is necessary. However, in OA with situs inversus totalis, left thoracotomy should be preferred.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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