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A case of esophageal atresia complicated by a right-sided aortic arch with right ductus arteriosus and inferior vena cava interruption with hemiazygos continuation

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ABSTRACT

During the repair of esophageal atresia with tracheoesophageal fistula (EA/TEF), cardiovascular malformations occasionally create a technical challenge. We report a novel case of a 4-day-old girl with EA/TEF and multiple cardiovascular malformations, including right-sided aortic arch (RAA), right ductus arteriosus (RDA), single ventricle with single atrium, common atrioventricular valve, pulmonary atresia, bilateral superior vena cava, and interruption of the inferior vena cava (IVC) with hemiazygos continuation. In this case, a right-sided approach would require the mobilization of the RAA and RDA, which were supplying the pulmonary blood flow due to pulmonary atresia. Alternatively, the left-sided approach would require the mobilization of the hemiazygos vein, which was essential for venous return from the lower body due to IVC interruption. We performed the less intrusive left-sided approach, and the postoperative course was uneventful. Right-sided EA/TEF repair should be avoided because RDA spasm or injury caused by RAA mobilization would be fatal. In cases of interrupted IVC with azygos or hemiazygos vein continuation, care must be taken not to ligate these vessels or block the venous return. Preoperative evaluation is important to prevent complications in such complicated cases. If sufficient information is not available, the left-sided approach may be preferred.

Congenital esophageal atresia (EA) is frequently associated with cardiovascular malformations [1]. These malformations sometimes make the surgical repair of EA remarkably challenging. This report describes a case of EA/tracheoesophageal fistula (TEF) associated with right-sided aortic arch (RAA) and right ductus arteriosus (RDA), which are cardiovascular malformations, thereby causing fatal complications during the EA/TEF repair with the right-sided approach.

1. Case report

A female infant was born after 37 weeks of gestation and weighed 2392 g. Ultrasonography revealed polysplenia and cardiovascular malformations, including RAA, single ventricle with single atrium, common atrioventricular valve, pulmonary atresia (PA), bilateral superior vena cava, and interruption of the inferior vena cava (IVC). A diagnosis of EA/TEF was confirmed by chest radiography. The stomach was present on her right side (Fig. 1). Because the esophageal gap measured three vertebral bodies in width and her general condition was stable, we planned a primary repair of the EA/TEF. A contrast-enhanced computed tomography (CT) was

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performed preoperatively to evaluate the surgical anatomy of the esophagus, trachea, and cardiovascular system. The CT revealed an RAA with mirror image branching, RDA, and interrupted IVC with hemiazygos continuation. Distal TEF was located between the descending aorta and the hemiazygos vein (Fig. 2a). The right-sided approach would require mobilization of the RAA and RDA, which provided the pulmonary blood flow due to PA (Fig. 2b). The left-sided approach would require mobilization of the enlarged hemiazygos vein (Fig. 2c).

Considering these findings, we performed the EA/TEF repair through left-sided thoracotomy on the fourth day of life. The hemiazygos vein was greatly enlarged and emptied into the atrium forming an extremely short arch on the dorsoventral axis, making access to the distal TEF difficult. We placed the loop around the hemiazygos vein and secured the surgical field by carefully handling the loop to not diminish the blood flow. After exposing the distal TEF behind the hemiazygos vein, the fistula was divided. The upper esophageal pouch was easily identified. The esophagus was anastomosed primarily by performing Livaditis myotomy. The postoperative course was uneventful, and milk intake was started 8 days after the surgery. On the 46th postoperative day, the left Blalock-Taussig shunt operation and RDA closure were performed by median sternotomy. The baby was discharged on the 57th day of life.

2. Discussion

EA/TEF is frequently associated with RAA [2,3], and discussion is ongoing regarding the surgical approach for the EA/TEF repair associated with RAA. Few authors recommended the left-sided approach [2–4], whereas others reported satisfactory results with the right-sided approach [5,6]. However, EA/TEF is often complicated by cardiovascular malformations in addition to RAA. Therefore, it is necessary to plan the surgery considering these complicated anatomical features.

Edwards [7] described three primary types of RAA. In each type, the ductus arteriosus may be on the left, right, or bilateral side (Table 1) [8]. In the case of RAA with left ductus arteriosus (LDA) (type IIIA1), the LDA runs from the left subclavian artery to the left main pulmonary artery. Therefore, injury to the LDA is unlikely during the EA/TEF repair performed using the right-sided approach. However, in type IIIA2, the RDA can be affected by the RAA mobilization during EA/TEF repair by the right-sided approach. It is established that the patent ductus arteriosus (PDA) is vulnerable and easily spasmed by stimulation. Even if the RDA was not directly injured, the spasm or flexion of the RDA could result in a loss of pulmonary blood flow, which might cause lethal consequences in cases of PA. This is the first case report of EA/TEF with RAA that explicitly mentions the presence of RDA. However, Knight et al. [9] reported that 13 out of 57 RAA cases had RDA. Thus, the probability of RDA is not low. In addition, Mentessidou et al. [4] reviewed 62 cases of EA/TEF with RAA and reported two deaths owing to intractable bleeding caused by RAA mobilization through the right thoracotomy and no deaths through the left thoracotomy. Although the PDA location was not described in either case leading to death [6,10], it is worth noting that the right-sided approach may cause fatal bleeding.

The azygos vein is often divided between ligations to gain complete exposure of the distal TEF in the EA/TEF repair. However, Antonio et al. [11] reported the case of IVC interruption with azygos vein continuation, wherein the interruption of venous drainage from the mid-lower part of the body after azygos division caused severe hypertension, acute renal failure, and IVC syndrome, ultimately resulting in death. Therefore, attention should be given to venous return in the case of an interrupted IVC. Our patient lacked the hepatic segment of the IVC, and the hemiazygos vein provided an alternative path to the single atrium. Because the hemiazygos

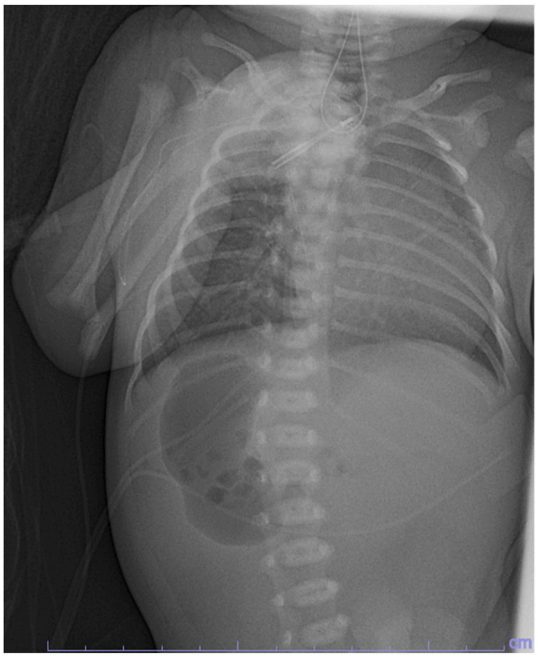

Fig. 1. Chest radiography taken after birth shows nasogastric tube folding in the upper pouch and right-sided stomach.

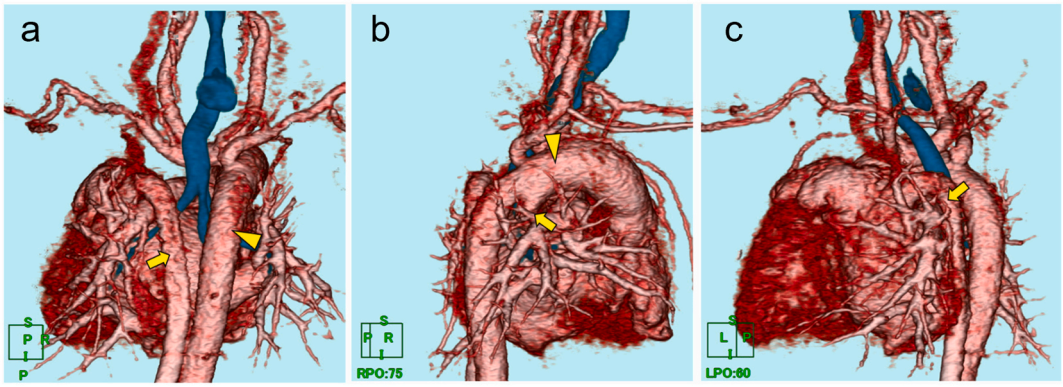

Fig. 2. Three-dimensional computed tomography (CT) image showing the location of the great vessels (red) and the trachea and esophagus (green).
a. Image taken from the dorsal side: The site of distal tracheoesophageal fistula (TEF) is located between the hemiazygos vein (arrow) and the descending aorta (arrowhead).
b. Image taken from the right side: The right aortic arch (arrowhead) and the right ductus arteriosus (arrow) are located anterior to the distal TEF.
c. Image taken from the left side: Hemiazygos vein (arrow) anterior to the distal TEF. . (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Table 1
Edwards' classification of the aortic arch [8].

I	Double aortic arch	A	both aortic arch patency	1: left ductus arteriosus
		B	one side aorta closed	2: right ductus arteriosus
				3. bilateral ductus arteriosus
II	Left aortic arch	A	normal branch	
		B	aberrant rt. subclavian artery	
		C	isolation of rt. subclavian artery	
III	Right aortic arch	A	mirror image branch	
		B	aberrant lt. subclavian artery	
		C	isolation of lt. subclavian artery	
IV	Others			

vein runs up to the left side of the vertebral column and anterior to the distal TEF, we had to mobilize the hemiazygos vein to expose the distal TEF without blocking venous return through the left-sided approach. It is worthwhile to recognize that the significantly extended hemiazygos vein makes this maneuver difficult.

Preoperative imaging is useful in these cases. Although ultrasonography is the most useful tool for evaluating cardiovascular anomalies, only 14%–80% of the RAA can be accurately identified preoperatively on ultrasound imaging [2,3,12]. Moreover, it is difficult to objectively evaluate the surgical anatomy of the trachea, esophagus, and cardiovascular system. Although the routine use of CT is not recommended owing to radiation exposure and sedation, it provides important information to avoid serious complications [13]. To select the appropriate surgical approach for treating EA/TEF in complicated cases, the following situations must be considered: First, it is necessary to classify the form of the aortic arch. We must remember that we can encounter RDA presenting anterior to the TEF through the right-sided approach in cases of RAA with RDA. Hence, in this case, or in cases where sufficient information about PDA is not available, the left-sided approach may be preferred. Second, the congenital anomalies of the systemic venous return should be confirmed. In cases of polysplenia, such as our case, interrupted IVCs are common [14]. The azygos vein should be preserved through the right-sided approach in the case of azygos continuation, while the hemiazygos vein should be treaded carefully through the left-sided approach in case of hemiazygos continuation. To avoid injury of these vessels, selecting the approach on the opposite side is preferable. Considering that the frequency of interrupted IVC with azygos continuation is higher than that in the case of hemiazygos continuation, left thoracotomy may also be safer if sufficient information about the systemic venous return is not available in the case of RAA. The limitation of our study is that it was a case report with only one patient. Further studies with a larger sample size are needed to determine the suitable approach for EA repair for each case.

3. Conclusion

In cases of EA/TEF with cardiovascular malformations, it is important to recognize the complicated anatomy. If sufficient preoperative information about the anatomy other than RAA is not available, then a left-sided approach may be preferable.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

Terutaka Tanimoto: data collection, written the paper.
Takuo Noda: study concept.
Hiroshi Nouso and Yukinori Miyata collaborated in the patient's medical care.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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