

# Right thoracotomy approach for patients with congenital tracheoesophageal fistula associated with right-sided aortic arch: a multicentric study

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**Background** A right-sided aortic arch (RAA) occurs in 5% of patients with esophageal atresia (EA). Its presence has significant surgical implications. Repair of the atresia has been considered difficult with the usual approach through the right chest. We hereby report our experience with cases of EA and RAA treated over the past 13 years in three pediatric surgical departments to determine the optimal surgical approach.

**Patients and methods** This is a retrospective study, which included all patients having EA with tracheoesophageal fistulas (TEFs) treated between January 2000 and September 2013 at four pediatric surgery departments (Zagazig University Hospital, Egypt; Assir Central Hospital, Kingdom of Saudi Arabia; MCH Najran, Kingdom of Saudi Arabia; Soba University Hospital, Sudan). The study compared a group of 22 patients who had EA with TEFs with RAA and no associated congenital disorder with another group of 22 patients who had the same condition but with normally situated left-sided aortic arch.

**Results** A total of 44 patients having EA with fistula were included into this study. They were divided into two groups on the basis of the side of aortic arch; 22 patients had left-sided aortic arch (group L), and the other 22 patients had RAA (group R). All patients were treated through right

thoracotomy. It was possible to achieve successful, although difficult management, in all patients with RAA through right thoracotomy. There was significant difference between the two groups of patients with regard to the operative time ( $104 \pm 9.8$  vs.  $149 \pm 15.4$  min). Twenty patients (45%) developed postoperative complications, with a morbidity rate of 41 and 50%, respectively, with no significant difference between the two groups. No mortality was recorded.

**Conclusion** Repair of congenital TEF associated with RAA can be performed through right thoracotomy safely but with some difficulty and longer time. *Ann Pediatr Surg* 11:120–122 © 2015 Annals of Pediatric Surgery.

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## Introduction

Esophageal atresia (EA), with or without tracheoesophageal fistula (TEF), is a fairly common congenital disorder that physicians should consider in the differential diagnosis of a neonate who develops feeding difficulties and respiratory distress during the first few days of life. Recognition of symptom complexes and a high index of clinical suspicion should assist early diagnosis [1–5].

Right-sided aortic arch (RAA) is a rare congenital anomaly, presenting in 0.1% of the general population. Embryologically, the aortic arch is formed from the left aortic arch (LAA) as the RAA is degenerated. However, if the LAA is degenerated instead of the RAA, RAA will be formed [6]. The RAA does not manifest symptoms necessarily, and it does not need surgical correlation. However, this condition is occasionally associated with other congenital diseases, including EA, TEF, vascular ring, and aortic arch anomaly around the trachea and esophagus, which can cause clinical symptoms such as respiratory obstruction and dysphagia [7].

RAA occurs in about 5% of infants with EA and TEF (EA/TEF). These patients have a high incidence of associated cardiac anomalies and occasional vascular rings [8,9]. The

preoperative diagnosis of patients with RAA is difficult. The preoperative diagnostic techniques may fail to show RAA associated with EA/TEF, and the diagnosis is often only made after right thoracotomy. A preoperative echocardiogram correctly identified the location of the arch in all patients with LAA but only in 62.5% of those with RAA [10–13]. The presence of a preoperatively unrecognized RAA makes the surgeon consider carefully whether or not to conduct the anastomosis through right thoracotomy [13,14]. It remains controversial whether right or left thoracotomy is the most appropriate surgical approach because primary repair of TEF is technically challenging in the presence of RAA [10,12,15,16].

In this retrospective study we present the experience of our centers in the management of cases of EA/TEF associated with RAA, which were discovered accidentally after right thoracotomy.

## Patients and methods

A total of 503 infants with EA/TEF were treated during the period from January 2000 to September 2013 at four pediatric and cardiothoracic surgery departments (Zagazig University Hospital, Egypt; Assir Central

Hospital, Kingdom of Saudi Arabia; MCH Najran, Kingdom of Saudi Arabia; Soba University Hospital, Sudan). All patients who had EA with TEF with RAA without associated congenital anomalies (22 out of 503) were included in our study and labeled as group R. This group was compared with a similar number of patients (22) who had EA with TEF with normally placed LAA and without any associated congenital anomalies (group L).

The diagnosis of EA/TEF was confirmed with a chest radiograph after passing a feeding tube into the upper esophageal pouch. The gap length was estimated on the basis of the position of the tip of the feeding tube and the apparent bifurcation of the trachea as seen on the chest radiograph. Absence of air in the gastrointestinal tract would raise concern of EA without fistula. Those patients without fistula were excluded from our study.

Careful intraoperative routine monitoring included an arterial catheter, ECG monitor, pulse oximeter, and a thermal probe. An awake intubation was commonly performed to allow spontaneous breathing. All patients underwent standard tracheal intubation with no attempt made to obtain single lung ventilation.

All cases were treated through right thoracotomy. At operation, once RAA was found, surgeons did not convert to left thoracotomy. The anterolateral portion of aortic arch was freed from the underlying distal esophagus and TEF; this allowed retraction of the aortic arch posteriorly, and the fistula was found under it and ligated. All cases were managed successfully through right thoracotomy.

Proximal pouch identification was facilitated with gentle pressure applied to the oral tube by the anesthesia team. A traction suture was placed to aid dissection of the proximal pouch. The proximal atretic esophagus was mobilized up to the level of the thoracic inlet and into the neck to acquire adequate length to achieve primary repair without tension. This was performed with gentle blunt dissection and care was taken to avoid injury to the trachea. Extensive dissection around the open distal

esophagus was avoided to reduce the risk of ischemia of mid-esophagus. Once adequate mobilization was achieved, a tension-free primary anastomosis was performed.

### Statistical analysis

The data were collected, organized, and tabulated, with particular reference to birth weight, gestational age, associated anomalies, preoperative investigations, operative approach and findings, perioperative complications, overall mortality, and long-term prognosis. Finally, the data were statistically analyzed using SPSS software statistical computer package, version 13 (IBM, New Orchard Road, New York). For quantitative data, the range, mean, and SD were calculated. For qualitative data, comparison between the two groups was made using the  $\chi^2$ -test. Correlation between variables was evaluated. Significance was adopted at *P* value less than 0.05 for interpretation of results of tests of significance.

This study was approved by the ethical committee of our institutions.

### Results

A total of 44 patients (100%) with EA/TEF were enrolled into this study. Of them, 25 (57%) were male and 19 (43%) were female patients. They were divided into two groups on the basis of the side of the aortic arch. Twenty-two patients (4.4%) had RAA (group R) and the other 22 patients had LAA (group L). We found no significant differences between the two groups with regard to patients' characteristics at admission (Table 1).

All patients were treated through right thoracotomy. Primary anastomosis was achieved successfully in all patients. A successful but difficult management was achieved in all patients with RAA through right thoracotomy. There was significant difference between the two groups of patients with regard to the operative time; otherwise there was no significant difference between the two groups in the operative and postoperative data (Table 2). Twenty patients (45%) developed postoperative complications, with no significant difference between the two groups. Seven patients developed

**Table 1 Preoperative data**

Characteristics	Group L (22 infants)	Group R (22 infants)	<i>P</i> value
The average age of mothers (mean $\pm$ SD) (years)	29 $\pm$ 3.8	31 $\pm$ 2.6	0.864
Gestational age (weeks)	36 $\pm$ 4 (27–41)	37 $\pm$ 3 (28–42)	0.352
Prematurity (<37 weeks) (%)	27%	30%	0.261
Birth weight (kg)	2500–4000 g (average, 2.8 kg)	2400–4100 g (average, 2.6 kg)	0.612
Male: female (%)	12:10	13:9	0.787
Delivery (vaginal delivery: cesarean section)	9:13	8:14	0.251

**Table 2 Operative and postoperative data**

Variables	Group L (22 infants)	Group R (22 infants)	<i>P</i> value
Operation performed			
Primary anastomosis	22	22	
The average length of the gap from proximal to distal oesophageal pouch (cm)	2.1 (range: 1.4–3.2)	2.2 (range: 1.5–3.4)	0.437
The average operative time (min)	104 $\pm$ 9.8	149 $\pm$ 15.4	0.031*
Average hospital stay (days)	23 $\pm$ 12	26 $\pm$ 14	0.263
Morbidity rate [ <i>n</i> (%)]	9 (41)	11 (50)	0.336

\*Significant (*P* < 0.05).

anastomotic strictures, three had anastomotic leakage, six had gastroesophageal reflux, and four had recurrent pneumonia. All complications were managed conservatively and the patients were discharged after improvement. No mortality was recorded.

## Discussion

Management of a child with EA/TEF with a RAA is technically challenging. Preoperative identification of the great vessel anomalies cannot be easily defined, and the surgeon may come across a RAA during surgery through right thoracotomy [13]. This study has reviewed our experience in the management of these anomalies.

The prevalence of male patients in our study was similar to that reported in other series [12]. The diagnosis of RAA was not suspected on preoperative chest radiography in any of our infants. Echocardiography was not performed routinely for our patients. Unfortunately, many authors documented that the diagnosis of RAA cannot be suspected with preoperative chest radiography in any infants and that echocardiography failed to diagnose RAA in most patients [11,15,16]. In one series, Allen *et al.* [10] cited that although preoperative echocardiogram could correctly identify the location of LAA, still RAA was detected only in 62.5% of their patients.

The incidence of RAA was found to be 4.4% in our study. The incidence was found to be 2.5, 3.7, and 13% by others [10,11,14]. In our results there were no significant differences between groups L and R as regards patient characteristics at admission. These results were nearly similar to that reported in other series [13].

In our series, although operating on patients with RAA through a right thoracotomy was difficult and took longer time, there was no need to convert to left thoracotomy in any of our patients. This is in agreement with the results of other authors who achieved primary repair in all patients with RAA through right thoracotomy without any problem [11,16]. Moreover, primary anastomosis was achieved successfully in all patients, with no significant difference between the two groups.

In our series, the overall morbidity rate was 45.5% and the mortality rate was 16%. This is much less compared with the overall morbidity rate (67%) and mortality rate (30%) reported in other studies, [17–21] mostly because patients who had associated cardiac lesions or long gap were excluded from our study. In addition, the site of aortic arch did not affect the morbidity or mortality rates in our patients; this coincides with that reported in other series [11,12].

## Conclusion

Surgical repair of EA is better to be performed from the opposite side of the aortic arch because it is less demanding and less time consuming. However, if the surgeon comes across an unexpected RAA after right

thoracotomy, the operation can be completed successfully from right side but with considerable amount of patience and high degree of technical expertise.

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### Conflicts of interest

There are no conflicts of interest.

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