Management of double aortic arch in children

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Abstract

Introduction: Double aortic arch (DAA) is a rare congenital malformation. Strategies for diagnosis and treatment are different. Reports of thoracoscopic treatment of DAA are rare. We report a description of our diagnostic and therapeutic approach and evaluate outcomes.

Material & Methods: We reviewed charts of infants with double aortic arch who were hospitalized in our department from 2008 to 2014. Age at symptom onset, clinical presentation, diagnostic modality, operative details and post operative outcomes were retrieved.

Results: We identified 7 patients presenting at a median age of 11,8 months (range: 2 months-27 months) and median weight of 10kg. Respiratory symptoms were present in all cases. Difficulties in feedings were present in 3 cases. Chest radiography, oesophagogram and Computed Tomography (CT) with three-dimentional (3D) reconstructions were performed for all patients. One patient had laryngo tracheoscopy and 3 patients echocardiography. The dominant branch was the right one in all cases. Associated cardiac anomaly was found in 1 case. Operative approach was thoracotomy in 3 cases and thoracoscopic video assisted surgery (VATS) in 4 cases. Median operative time was 132 min. Only one patient required conversion. Median hospital stay was 4,1 days.

Conclusion: Double aortic arch is a rare malformation which can cause persistent non specific respiratory symptoms in infants. The use of 3D-CT scan reconstruction facilitates diagnosis and the operative approach. Thoracoscopy and thoracotomy are effective procedures even for patients with low body weight. The operative time seems similar for both approaches. Thoracoscopy offers less post-operative analgesia requirement, shorter hospital stay and good cosmetic results.

Keywords

- double aortic arch
- vascular ring
- surgery
- thoracoscopy
- complications
- child

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Introduction

Abnormalities of the aortic arches are rare malformations representing less than 1% of all congenital heart malformations. Double aortic arch (DAA) is categorized in complete vascular ring disorders.[1] Most articles referring to this topic are published as case reports. Diagnostic methods have evolved from chest radiography, esophagocardigram, echocardiography to computed tomography scan (CT) and magnetic resonance imaging (MRI).[2-4] Gross was the first surgeon who succeeded in the surgical repair of DAA in 1945 implicating the use of thoracotomy.[2] The first video assisted thoracoscopic division of vascular ring was accomplished recently and was described by Burke and Chang in 1993.[5] Since then, only small series using thoracoscopy are found in the literature and in our knowledge, no comparative studies between the two surgical techniques were published. We report methods of diagnosis and evaluate the efficacy of open and thoracoscopic treatment

Materials and methods

From January 2008 to December 2014, seven patients were hospitalized for DAA repair in the department of pediatric surgery at Fattouma Bourguiba Hospital. We reviewed their medical records with regards to age and weight at presentation, initial clinical features, diagnostic modality, operative details and post-operative outcomes.

All patients were symptomatic at the time of surgery. All patients were evaluated by esophagogram and CT scans with three dimensional (3D) reconstructions. Echocardiography was done in 2 patients and laryngo tracheoscopy in one case.

Thoracotomy was performed in 3 patients in 2008 and then, subsequently, video-assisted thoracoscopic surgery (VATS) was used for the other patients. VATS was done with patients in a lateral decubitus position. Three trocars were used. One 5mm was placed in the 5th intercostal space (ICS) in the mid-axillary line to accommodate the 30 degree telescope. Two additional ports were used, a 5mm trocar in the 4th ICS at the anterior axillary line and a 5mm trocar in the 6th ICS at the posterior axillary line. Left or right thoracoscopic approach was performed according to the side of the dominant arch. The procedure started by opening the mediastinal pleura, dissection of the ring elements and before division of the complete vascular ring we used a clamp test for the hypoplastic arch under pressure monitoring and we checked the carotid pulses. The section of this one is done after ligation of the arc by Nylon 2/0 allowing the oesotracheal decompression. All patients had chest tube at the end of the procedure under vision.

This study was approved by the ethics committee of the hospital.

Results

Over the seven year period, 7 patients were identified with the diagnosis of DAA. There were 4 boys and 3 girls. The mean age at diagnosis was 11.8 month (range: 2 months- 27 months). None of the patients had antenatal diagnosis of DAA. The mean weight was 10kg (range: 4kg-18kg). All patients were symptomatic at the presentation; mainly respiratory symptoms. The most common symptom was recurrent bronchitis Table 1.

Table 1: characteristics of patients with DAA

No	Age (months)	Weight (kg)	sex	symptoms	explorations	Dominant arch
1	11	11	male	Bronchitis + stridor + difficulties in feeding	LT CR+E ETT CT	right
2	5	7	female	Recurrent bronchitis	CR+E CT	right
3	12	9	male	Recurrent bronchitis	CR+E CT	right
4	3	5	male	Respiratory distress + difficulties in feeding	CR+E ETT CT	right
5	2	4	female	Stridor + Respiratory distress + difficulties in feeding	CR+E CT+ETT	right
6	23	16	male	Recurrent bronchitis	CR+E CT	right
7	27	18	female	Recurrent bronchitis + stridor	CR+E CT	right

LT: laryngotrecheoscopy, CR: chest radiography, E: esophagogram, ETT: trans-thoracic-echocardigraphy, CT: Computed tomography

All patients had chest radiography and upper gastrointestinal series showing in all cases a permanent imprint in the thoracic esophagus level suggesting a vascular ring anomaly Figure 1.

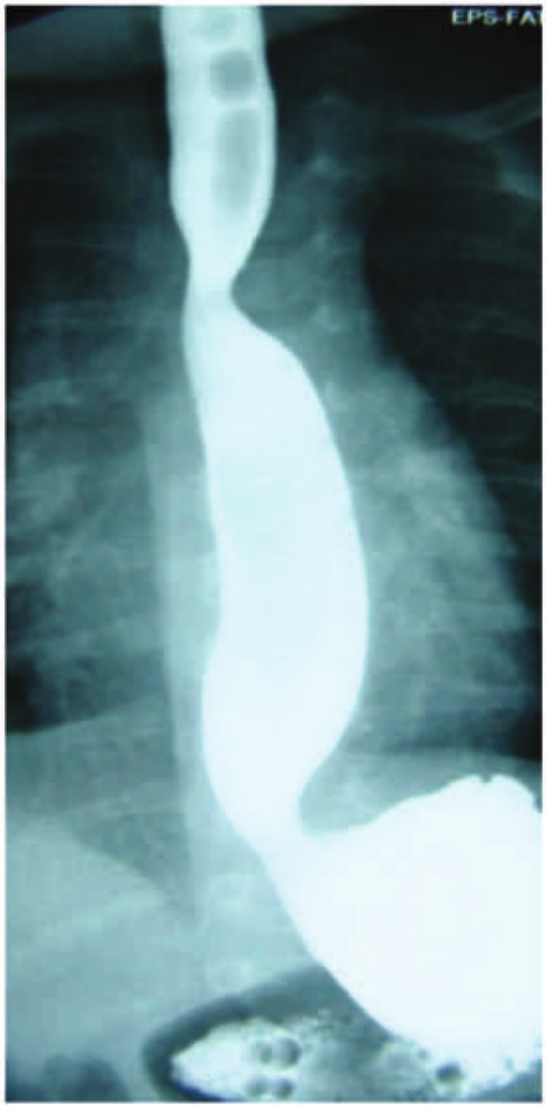

Figure 1: oesopagogram showing an imprint in the oesophageal wall evocating vascular ring anomaly

Echocardigraphy was performed in three cases and didn't lead to the diagnosis but in one case it showed a 5 mm associated inter auricular communication. One patient had laryngo tracheoscopy that revealed external pulsatile compression on trachea.

CT scan with three-dimentional (3D) reconstructions was performed for all patients making the diagnosis in all cases (Figure 2, 3) and helping in planning surgical strategy. The dominant branch of double aortic arch was the right one in all cases.

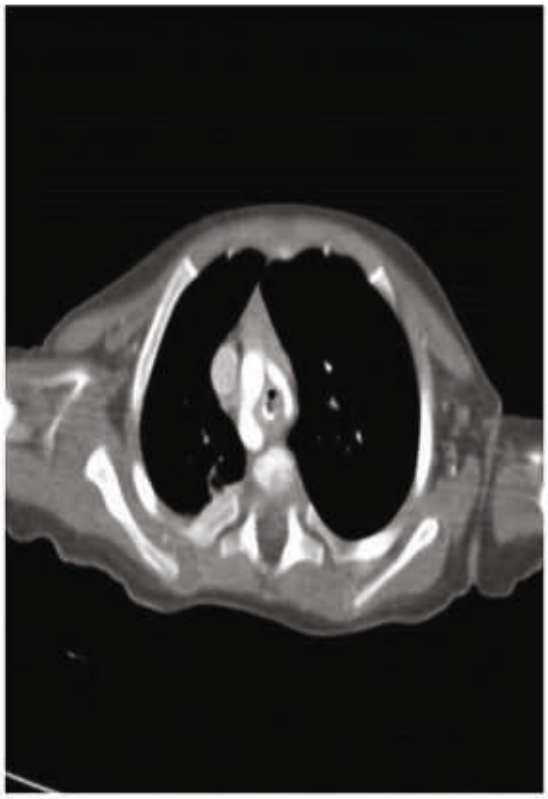

Figure 2: CT scan showing a double aortic arch with a right dominant arch

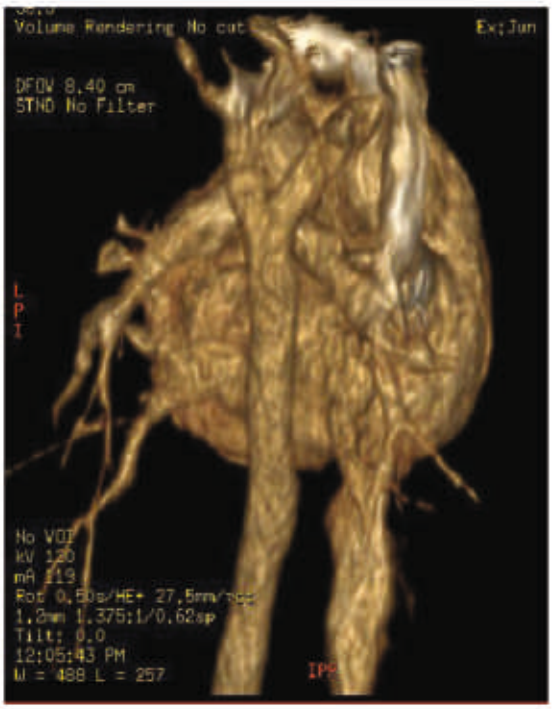

Figure 3: 3D CT scan reconstruction CT showing DAA

Operative approach was thoracotomy in 3 cases and thoracoscopic video assisted surgery (VATS) in 4 cases (Figure 4).

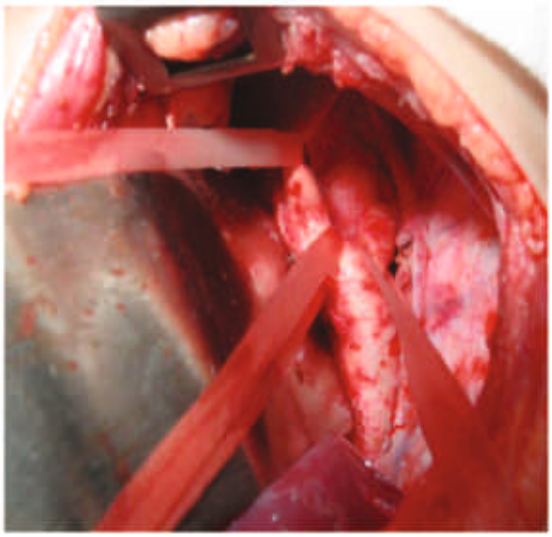

Figure 4: Intraoperative photo with individualization of the descending aorta and the left anterior arch.

Median operative time was 132 min (136.7 min for patients operated by thoracotomy vs 128.8 min for patients operated by thoracoscopy). Only one patient operated initially by thoracoscopy required conversion. In fact, the chest tube revealed permanent hemorrhage thus a thoracotomy was performed. There was a continuous bleeding from the sectioned arch. Ligation of the vessel was performed with good results and recovery. Median Length of analgesia was 3 days (2 days for thoracoscopy vs. 5 days for thoracotomy). Median hospital stay was 4.1 days (shorter when thoracoscopy). There was no late evidence of aortic arch obstruction at follow up Table 2.

Table 2: type of surgery and postoperative outcomes

No	Surgical approach	Duration (min)	Post operative analgesia (days)	Hospital stay (days)	complication	Follow-up (months)
1	Thoracotomy	125	5	5	no	66
2	VATS	105	2	3	no	62
3	VATS	130	2	3	no	30
4	VATS	160	5	5	Hemorrage-> conversion	18
5	Thoracotomy	150	5	5	no	68
6	VATS	120	2	3	no	6
7	Thoracotomy	135	5	5	no	68

Discussion

DAA is a congenital anomaly of embryonic aortic development, due to the persistence of the fourth right and left arches and dorsal aortas. It results in the abnormal formation of a complete vascular ring around trachea and esophagus.[6] The principal branch of DAA is frequently right. [6,7] In our study all patients had right predominant arch. Usually DAA occurs without associated cardiovascular malformations. However, anomalies like absent left pulmonary artery, tetralogy of fallot and esophageal atresia[7-9] have been mentioned as associated anomalies. Rare syndromic malformations such as VACTERL, Prader-Willi [10] and Pierre-Robin[11] were also reported as associated anomaly and should be sought.

Prenatal diagnosis is possible.[12, 13] In our study, antenatal ultrasonography didn't show any abnormality. After birth, aortic arch abnormalities should be considered as a differential diagnosis in infants with recurrent respiratory signs (bronchitis, dyspnea, stridor, respiratory distress, apnea, and cyanosis).[6-9] Gastro intestinal manifestations may also be present, such as dysphagia and difficulties with feedings. In our study,

respiratory symptoms were predominant.

Diagnosis is often difficult and may be missed. AL-Bassam et al[1] and Ruzmetov et al[7] found that esophagogram is an excellent mean for demonstrating the presence of a vascular ring. It can also demonstrate an associated esophageal reflux. In all our cases we found a persistent imprint on the esophageal wall. Echocardiography may confirm associated cardiac malformations yet a DAA abnormality may be missed.[3, 10] As the patients typically present with respiratory symptoms, bronchoscopy can be asked as an initial investigation. It showed in one of our cases an external pulsatile compression on trachea. However, it cannot always specify the anatomical type of vascular ring. It may also be responsible for edema which aggravates tracheal obstruction.[10] In our study CT scan with 3D reconstruction functioned perfectly for diagnosis of DAA preoperatively. It provided complete information regarding the vessels responsible for tracheo-esophageal compression; thus, helping to choose the appropriate approach to best deal with the hypoplastic arch. Multi-slice CT scan offers several advantages[3&4]

compared to MRI in selected patients. MRI has been shown to accurately depict the level, severity and cause of vascular compression without the drawback of using ionizing radiation in the process. However, MRI has the disadvantage taking considerably longer time to operate. Additionally, it is a costly modality, and is often more difficult to schedule than CT.[4]

Surgical treatment of DAA mainly consists of a left or right thoracotomy depending on the side of the dominant arch.[2] Post-operative complications are moderately rare. Alsnaidi et al encountered post operative complications in 9% of their 81 cases of DAA.[6] The rate of complications with Bonnard et al, was 12% in 62 patients operated for vascular ring abnormalities. Reported complications were hemorrage, chylothorax, pneumothorax and sepsis.[9] For Ruzmetov et al[7], post-operative complications occurred in 2% of their cases. No patient died in his study at follow up because of post operative complication. Alsnaidi et al[6] reported 2 deaths after surgical repair of DAA with an estimate survival of 96% at 5 years. According to him, persistent respiratory symptoms are associated with previous compression related to abnormal development of the trachea and airways.

VATS is an alternative to open thoracotomy for management of DAA. In 1993, Burke and Chang[5] reported the first case of thoracoscopic division of vascular ring. The number of patients especially infants operated subsequently is very small, often reported as case reports. The series published regarding VATS for DAA in literature are usually associated with other vascualr ring abnormalities repares.[1,14] Burke et al, reported a series of six infants and children with vascular ring.[14] Al Bassem et al[1], reported only a series of 4 patients operated for right aortic arch. None of them needed a second operation and complete resolution of symptoms was noted. In our knowledge, no comparative studies between open and thoracoscopic surgery of DAA were published.

Although our series is too small to give statically significant results, it seems that thoracoscopy and thoracotomy are effective procedures for the treatment of DAA even for patients with low weight. The operative time seems similar for both methods. Patients treated with thoracoscopy need less post-operative analgesia, have shorter hospital stay and good cosmetic results. However, it needs a trained surgeon experienced in open surgery when conversion is indicated. A Study with a bigger number of patients is desirable.

Reference:

1. Al-Bassam A, Saquib Mallick M, Al-Qahtani A et al: Thoracoscopic division of vascular rings in infants and children. J Pediatr Surg. 2007; 42(8):1357–1361.
2. Gross RE: Surgical relief for tracheal obstruction from a vascular ring. N Engl J Med 1945;233: 586.
3. Hetal N Jeeyani, Vaishali J Prajapati, Nehal H Patel et al : Imaging features of double aortic arch shown by multi detector computed tomography angiography. Ann Pediatr Cardiol. 2010; 3(2): 169–170.
4. Cerillo AG, Amoretti F, Moschetti R et al: Sixteen-row multislice computed tomography in infants with double aortic arch. Int J Cardiol. 2005; 99(2):191–4.
5. Burke RP, Chang A: Video-assisted thoracoscopic division of a vascular ring in an infant: a new operative technique. J Card Surg. 1993; 8: 537 - 40.
6. Alsenaidi K, Gurofsky R, Karamlou T, et al: Management and outcomes of double aortic arch in 81 patients. Pediatrics. 2006; 118(5): 1336-41.
7. Ruzmetov M, Vijay P, Rodefeld MD et al: Follow-up of surgical correction of aortic arch anomalies causing tracheoesophageal compression: a 38-year single institution experience. J Pediatr Surg. 2009; 44:1328–1332
8. Majid Y, Warade M, Aziz Z et al: Double aortic arches, esophageal atresia and tracheal compression. J Indian Assoc Pediatr Surg 2009;14(2):70-2.
9. Bonnard A, Auber F, Fourcade L et al: Vascular rings abnormalities: a retrospective study of 62 cases. J pediatr surg 2003; 38(4):539-43.
10. Shanmugam G, Macarthur K, Pollock J. Surgical Repair of Double Aortic Arch: 16-year Experience. Asian Cardiovasc Thorac Ann. 2005;13:4-10.
11. Waters ET, Oberman JP, Biswas AK: Pierre Robin sequence and double aortic arch: a case report. Int J Pediatr Otorhinolaryngol. 2005;69:105-10.
12. Sánchez Andrés A, Arenas Ramirez J, Rodriguez Fernández A: Anillo vascular completo: diagnóstico prenatal de doble arco aórtico. An Pediatr (Barc). 2014;81(4):260-2.
13. Tuo G, Volpe P, Bava GL, et al: Prenatal diagnosis and outcome of isolated vascular rings. Am J Cardiol. 2009;103(3):416-9.
14. Burke RP, Rosenfeld HM, Wernovsky G, et al: Video-assisted thoracoscopic division of vascular rings in 6 infants and children. J Am Coll Cardiol 1995;25:943 - 7.