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How Far Aortopexy Favors the Outcome of The Doubly-Aortic-Arched Patients

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ABSTRACT

Background: Treatment for various vascular ring malformations remains difficult due to the fact that they need distinct surgical techniques. In this retrospective analysis, we offered our four-year experience with surgical therapy in vascular rings.

Objective: We aimed at determining the impact of aortopexy technique in patients with double aortic arch (DAA) on early extubation and improvement in both early and late-term outcomes.

Patients and methods: The study involved 30 patients who had surgical therapy for DAA between January 2021 and January 2024. They were divided into two groups: Group (A) included 15 patients who were repaired with aortopexy technique and group (B) included 15 patients who were repaired without aortopexy technique.

Results: The patients had a mean age of 41.50 ± 38.61 months and a mean weight of 42.00 ± 63.47 kg at surgery. The most prevalent symptom was a cough. Shortness of breath, trouble swallowing, reflux, and recurring pneumonia follow. We encountered early problems such as cylothorax, pneumothorax, and the requirement for a tracheostomy (due to failure of extubation) in 5 patients (16.66%). Patients experienced no long-term problems, with the exception of one (3.33%) incidence of asthma. However, no pressure was recorded on the patient's trachea. We had no fatality during hospitalizations or the follow-up period. The mean duration of mechanical ventilation (MV) in group (A) patients was 3.10 ± 16.79 hours and 10.00 ± 16.79 hours in group (B) patients ($p< 0.001$).

Conclusion: Vascular rings can appear at different ages and with varied symptoms. But, once the diagnosis was established, surgery was necessary and produced acceptable, uneventful long-term survival outcomes. Aortopexy with repair of DAA was beneficial for early extubation and improvement of tracheomalacia.

Keywords: Vascular rings, DAA, Aortopexy.

INTRODUCTION

In cases of vascular rings, the trachea and/or esophagus are compressed during the embryonic period due to aberrant development of the aortic arch and its branches. This can lead to symptoms like coughing, recurrent respiratory infections, wheezing, dysphagia, or respiratory distress. Steroid wheeze and stridor are the most common [(1)].

Less than 1% of congenital cardiac abnormalities are vascular rings. The disease's forms are primarily classified into two groups based on whether the trachea and esophagus are completely or partially encircled. The right aortic arch with left ligamentum and DAA are the two most prevalent vascular rings types [(2)]. Nonetheless, depending on the severity of the tracheo-esophageal pressure, the recurring and worsening clinical signs linked to vascular rings may necessitate immediate care and demand a thorough examination. Numerous imaging and endoscopic techniques, including CT, MRI, bronchoscopy, gastroscopy, echocardiography, and cardiac catheterization, can be used to confirm the diagnosis [(3)].

Following diagnosis, typical therapy for decompression calls for surgical intervention. Although surgical approaches vary based on the anatomical aspects of the illness, several reports showed good long-term outcomes with minimal mortality and no events. However, despite the procedure, the respiratory issue could persist because of a structural deficiency and acquired tracheomalacia brought on by persistent pressure on the airways [(4)].

This study primarily aimed at assessment of the impact of aortopexy technique in patients with double aortic arch on early extubation. Secondary endpoint was to determine its impact on the improvement in both early and late-term outcomes.

PATIENTS AND METHODS

Study design: This retrospective study included 30 symptomatic patients with the diagnosis of vascular ring (double aortic arch). They were divided into two groups: Group (A) included patients that were repaired with aortopexy technique including 15 patients and group (B) included 15 patients that were repaired without aortopexy technique including. All surgeries were performed in Egypt (in the operating rooms of the Department of Cardiothoracic Surgery, Faculty of Medicine, Cairo University, and Beni-Suef University) with conventional open-heart surgical procedures.

The study's data were collected for individuals who had been operated on between January 2021 and January 2024. All data were collected and extensively examined before to, during, and after surgery.

Inclusion criteria: Those diagnosed with isolated vascular ring (double aortic arch).

Exclusion criteria: Patients with other congenital cardiac defect requiring repair or surgical intervention at the same setting.

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Management protocol:

(1) Preoperatively:
The preoperative variables that were assessed included: Demographic data of the patients, preoperative symptoms such as cough, dyspnea, dysphagia, wheezing, stridor, or repeated respiratory infections, and imaging methods used to confirm the diagnosis: transthoracic echocardiography (TTE), computed thorax angiography (CTA), magnetic resonance angiography (MRA), bronchoscopy, and barium esophagography.

(2) Intraoperatively:
The operative variables that were analyzed included intraoperative mortality, operative technique and inotropic support demand.

(3) Operative Technique:
The fourth intercostal gap was used to reach the pleural cavity in DAA patients after a posterolateral thoracotomy incision. The tiny aortic arch was split after both were evaluated. At first, the arches' diameters were assessed. The hypoplastic one was divided, which decompressed the esophagus and trachea. When the two diameters were comparable, the limbs' blood pressure was assessed, and the arch on the side where the lower arterial pressure was found was then separated. Additionally, the right arch division was selected because it circumscribes the trachea and exhibits equality in blood pressure measurement and diameter size. Recompression was prevented, nonetheless, since the descending portion of the split arch was stabilized at the thorax wall using polypropylene sutures. The pleura in the middle was always maintained open. We elevated the aorta anteriorly and sutured it to the posterior aspect of the sternum in aortopexy patients. Aortopexy opens the tracheal lumen because the anterior tracheal wall is connected to the posterior aorta wall through the pre-tracheal fascia.

(4) Postoperatively:
The postoperative variables that were assessed included hemodynamic status in the ICU, duration of MV, duration of inotropic support, total ICU stay, immediate postoperative mortality, morbidity, adverse complications during hospital stay and total duration of hospital stay.

Ethical Approval: Research Ethics Committee of The Cardiothoracic Surgery Operating Theaters, Cairo University and Beni-Suef University approved this study with approval number of FMBSUREC/07042024/Elbatanony. To participate in the study, each patient signed a written informed consent form. The Helsinki Declaration was adhered to at every stage of the study.

Statistical analysis

Using SPSS version 21.0, the gathered data were arranged, tabulated, and statistically examined. The X^2-test or, when applicable, Fischer's exact test were used to compute the frequency and percent distributions for the qualitative data. In the case of quantitative data, the t-student test was used to compare the mean ± SD, minimum, and maximum values. A p-value ≤ 0.05 was deemed significant in all tests.

RESULTS

The whole study population composed of 30 patients. 8 (26.66%) were females and 22 (73.33%) were males. Their mean age was 41.50 ± 38.61 months. Their preoperative mean weight was 42.00 ± 63.47 kg. There were no associated anomalies and all of them were operated for isolated DAA anomalies. The most common symptom among patients was cough. Dyspnea and dysphagia were following respectively. Besides, acid reflux, cyanosis and recurrent respiratory infections were less frequent. All patients were examined via TTE. The definitive diagnosis was made by MRA in 3 (10%) patients and CTA in 13 (43.33%) patients (Table 1).

Table (1): Preoperative characteristics.

Characteristics	
Age (months)	41.50 ± 38.61
Gender (Females/Males) (%)	8/22 (26.66/73.33)
Weight (kg)	42.00 ± 63.47
Cough (%)	13 (43.33)
Dyspnea (%)	10 (33.33)
Dysphagia (%)	3 (10)
Acid Reflux (%)	2 (6.66)
Recurrent chest infections (%)	1 (3.33)
Cyanosis (%)	1 (3.33)

Because of the presence of tracheomalacia, bronchoscopy and barium esophagography were performed on one (3.33%) patient each. In group (A), the average length of MV, ICU stay, and hospital stay were 3.10 ± 16.79 hours, 40.50 ± 26.33 hours, and 5.00 ± 2.62 days, respectively. In group (B), the average length of MV, ICU stay, and hospital stay was 10.00 ± 16.79, 45.50 ± 26.33 hours, and 7.00 ± 2.62 days respectively. Three (20%) individuals in group (A) had early problems. These included pneumothorax, chylothorax, and tracheostomy placement (due to failure of extubation in one (6.66%) patient with preoperative tracheomalacia). In group (B), two patients (13.33%) had early problems. These were chylothorax and tracheostomy application (due to failure of extubation in one (6.66%) patient who had tracheomalacia preoperatively). There was no intraoperative or in-hospital postoperative mortalities. Early postoperative complications and ICU parameters are illustrated in table (2). The cumulative duration of the study was 4.00 years. There were no mortalities during the follow-up period.

Table (2): Early postoperative complications and ICU parameters.

	Group (A)	Group (B)	*p* value
Mean duration of Mechanical Ventilation (hours)	3.10 ± 16.79	10.00 ± 16.79	**<0.001**
Mean ICU stay (hours)	40.50 ± 26.33	45.50 ± 26.33	0.855
Mean hospital stay (days)	5.00 ± 2.62	7.00 ± 2.62	0.741
Pneumothorax (%)	1 (6.66)	0	0.101
Chylothorax (%)	1 (6.66)	1 (6.66)	-
Failure of extubation (%)	1 (6.66)	1 (6.66)	-

DISCUSSION

Treatment remains difficult because various surgical techniques are needed for distinct vascular ring abnormalities [5]. Furthermore, depending on the degree of compression over the trachea and/or esophagus, the presentation of vascular rings can range from severe respiratory issues that arise in the neonatal period to non-specific findings at school age [5, 6].

In some situations, the diagnosis could go unnoticed and only be discovered by chance when looking at fewer common symptoms [7, 8, 9]. For instance, the asthma and dyspnea brought on by exercise that our sample experienced. Furthermore, during our search of the literature, **Sturm *et al.*** [10] reported on a child who was eight years old and receiving treatment for asthma during a five-years' period.

In our cohort, at the time of diagnosis, patients ranged in age from 4 to 132 months and displayed a wide spectrum of signs and symptoms. The most typical symptom was cough, which was followed by dysphagia, acid reflux, recurrent pneumonia, and shortness of breath. When these recurrent songs occur, the physician should be informed about possible vascular rings.

These days, non-invasive procedures like TTE, CTA, and MRA have taken the place of formerly popular diagnostic approaches [11]. Nevertheless, bronchoscopy, esophagoscopy, and barium swallow x-ray were necessary for the diagnosis in 3 (or 10%) of our patients. Every patient should have a TTE examination in order to check for concomitant cardiac abnormalities, regardless of the method used for the patient's diagnosis. TTE may not always be successful, in which case more sophisticated techniques may be required. While, angiography and cardiac catheterization were the most common procedures used in the past, CTA and MRA are currently preferred [6, 8, 9, 11]. There are six primary groupings under which vascular rings are categorized: The development of the right-sided aortic arch plus aberrant left subclavian artery as a result of the aortic arch's persistence, the left aortic arch plus aberrant right subclavian artery following the regression of the fourth right arch between the carotid and subclavian arteries, and the persistence of the fourth right aortic arch all contribute to DAA. There was just one of them in our series. In 1999, **Backer *et al.*** [1] made revisions to this categorization.

Determining the dominant arch requires a thorough evaluation prior to surgical correction [12].

It is advised that surgery be carried out via the dominant arch's opposing side [13]. The posterolateral thoracotomy with muscle sparing was our favored method. The ligamentum arteriosum, the subclavian artery, and the distal arch must all be identified by meticulous dissection after accessing the thoracic space through the fourth intercostal gap, lung retraction, and opening the covering pleura on the vascular ring. Surgeons should be very aware of and cautious throughout this procedure to prevent damage to the thoracic duct, phrenic, vagus, and recurrent laryngeal nerves, especially in the left chest. Nonetheless, blood pressure testing is necessary to determine which arch will be dissected when both have equal diameter sizes [14]. Another significant step is to conclude surgery with the mediastinal pleura left open [1, 2].

According to **Backer *et al.*** [15]**,** during ring division, surgeons may contribute to the development of recurrent scar tissue and recurrent stenosis. In other words, it is possible to prevent any compression on the trachea and esophagus brought on by bleeding sites or fluid.

For the division of vascular rings, several facilities choose for video-assisted thoracoscopic surgery (VATS) [16]. Despite lacking prior expertise in VATS surgery, **Riggle *et al.*** [17] indicated that VATS is a realistic procedure that may be conducted with good outcomes. There was a reduction in the duration of the operation, ICU admissions, chest tube usage, chylothorax, and total hospital stay. However, given the location of the arches beneath the esophagus and the potential risk of significant bleeding, **Backer *et al.*** [15] highlighted that this risk may not be controllable by a clip or staple line slide off a vessel. Furthermore, they recommended remembering that the atretic arch possesses a little patent lumen.

In addition to bleeding, vascular ring surgical problems have been reported including neuralgia, Horner's syndrome, pneumonia, pneumothorax, chylothorax, and vocal cord paralysis [1, 2, 8, 9, 15]. Three instances (20%) were seen; one case (6.66%) involved chylothorax, one case (6.66%) involved pneumothorax, and one case (6.66%) involved the application of tracheostomy due to extubation failure in a patient who had been preoperatively diagnosed with tracheomalacia. The range of complications rates reported in the literature is 10-27%. Nonetheless, recorded death rates are less than 1% [15, 16, 18]. Additionally, none of the 30 patients that we had during their in hospital stay or follow-up period died.

In most situations, surgical therapy results in an uneventful survival. In their late follow-up, **Backer *et al.*** [15] found that 92% of the cases had progressed without any symptoms. Additionally, out of 350 patients, only 8 instances were recorded requiring re-intervention after Kommerell diverticulum was eliminated. Other researchers observed similar results [18, 19]. We carried out an aortopexy in our series. By doing this, we reported a statistically significant difference in early postoperative extubation and less persistent late-term respiratory problems. The mean duration of MV was 3.10 ± 16.79 hours for patients in group (A) and 10.00 ± 16.79 hours for patients in group (B) in our research ($p<0.001$).

Long-term, substantial tracheal compression can result in a localized region of tracheomalacia, irreparable damage to the tracheal structure, and chronic discomfort. About 70–75% of symptoms go away in the first few days after surgery. About thirty percent of the symptoms linger, according to some writers, and they usually go away later [12, 15, 16, 19, 20]. Aside from one (3.33%) episode of asthma, we also did not have any lingering effects. On the trachea, no appreciable pressure was found in this patient, nevertheless.

Study Limitations: The current study had limitations due to its retrospective nature. Furthermore, the small number of patients and lack of data on some forms of vascular ring categorization appear to be distinct limitations.

CONCLUSION

Vascular rings can appear at any age and with a variety of symptoms, therefore diagnosis is dependent on awareness. Surgical intervention is necessary as soon as the patient has been diagnosed in order to stop any permanent damage. When deciding how best to do the procedure, a thorough examination and appraisal of the anatomy are helpful. The purpose of the procedure is to divide the displaced artery and maintain enough blood flow to the head in order to alleviate compression on the trachea and/or esophagus. Following the procedure, the results are good and result in a smooth long-term survival. In our study, it was beneficial to do aortopexy for cases with DAA repair as there was a significantly obvious improvement of tracheomalacia and early extubation.

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