Case Series

Partial upper median sternotomy for anterior aortopexy for innominate artery compression syndrome: a case series

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Background: Innominate artery compression syndrome (IAS) is caused by an abnormally originating innominate artery compressing the trachea anteriorly. One option to relieve such compression is an anterior aortopexy (AA). We describe our technique of an AA via a partial upper median sternotomy.

Case Description: Nine consecutive patients underwent AA for IAS via a partial upper median sternotomy from July 2017 to November 2020 at two US teaching hospitals. The median age was 9 months [interquartile range (IQR), 3–16.5 months]. The male to female ratio was 1.25. All patients had >70% compression by flexible bronchoscopy. Two patients had previous surgeries. The median follow-up was 6 months (IQR, 4–8.5 months). The indications for the operation were: acute life-threatening events (ALTEs) (4/9 patients), recurrent intubation (4/9), and severe stridor with >70% luminal reduction (1/9). Technical success (defined as ≤20% residual stenosis) was achieved in 78% (7/9) of the patients. The two patients with unsuccessful AAs required either a tracheal resection or an innominate artery reimplantation. Both achieved full symptom resolution. Overall, 78% (7/9) of patients experienced full symptom resolution. Of the two patients without full symptom resolution, one had mild stridor at 6 months post-operation. The other patient without full resolution is awaiting further vocal cord surgery for an associated glottic pathology.

Conclusions: A partial upper sternotomy provides a very versatile approach to an AA for IAS. In addition to facilitating an adequate AA, a partial upper sternotomy provides options for direct tracheal surgery or an innominate artery reimplantation in case an optimal result is not obtained by an AA.

Keywords: Aortopexy; innominate artery compression syndrome (IAS); partial sternotomy; case series

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Introduction

Anterior aortopexy (AA) for innominate artery compression syndrome (IAS) was first described by Gross *et al.* in 1948 (1). IAS is thought to be due to an abnormal innominate artery arising distally along the aortic arch and crossing the trachea anteriorly from a left to right direction and thereby compressing the trachea. The tracheal compression can be exacerbated in younger children who have large thymuses and overcrowded upper mediastina (2,3).

Common symptoms of IAS include stridor, acute life-threatening event (ALTE), and recurrent respiratory infections. IAS can also present as failure to wean from mechanical ventilation. Indications for surgery include ALTE, two or more episodes of infection, >70% luminal reduction, and dependence on mechanical ventilation. ALTE is the strongest indication for an intervention as it

can lead to mortality (4-6).

IAS is diagnosed by flexible bronchoscopy, which shows an anterior pulsatile compression in the distal trachea. Flexible bronchoscopy also guides the AA operation, helping to determine the number and position of anchoring sutures, and evaluating the result of the AA (*Figure 1*). Cross-sectional imaging is also obtained to further evaluate the anatomy.

Several approaches to perform an AA have been described, including a left anterior thoracotomy, right thoracotomy, median sternotomy, and thoracoscopic approaches (7,8). A partial upper sternotomy approach for AA for tracheomalacia from all causes has been described by Elliott *et al.* (8). This approach has become our preferred approach for an AA for IAS. Our objective is to illustrate our technique and describe the experience gained from nine consecutive cases using an upper partial median sternotomy approach to an AA specifically for an IAS. We present this case series in accordance with the AME Case Series reporting checklist (available at https://tp.amegroups.com/article/view/10.21037/tp-23-597/rc).

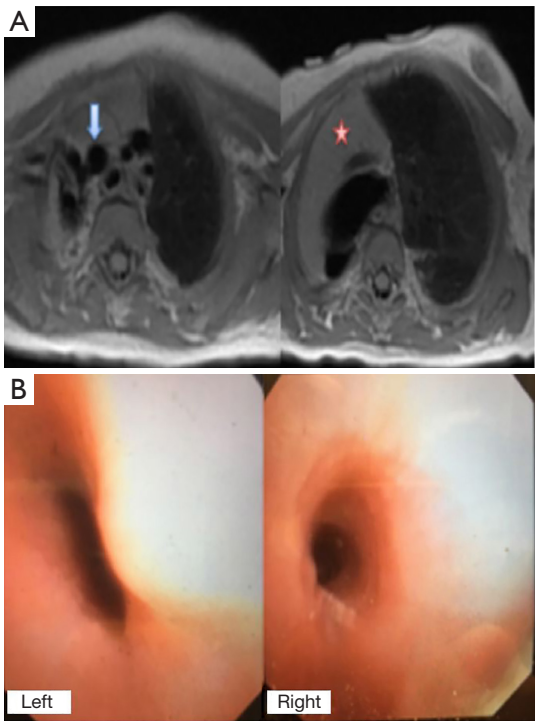

Figure 1 MRI and bronchoscopy. (A) A transverse section MRI scan of the chest showing the innominate artery (arrow) in close apposition to the trachea and causing compression. Also, note the large thymus (star) in the anterosuperior mediastinum causing crowding. (B) Flexible bronchoscopy showing: Left: anterior extramural compression of the trachea by the innominate artery causing >70% reduction if the luminal area. Right: 5% residual tracheal luminal area after a successful AA. MRI, magnetic resonance imaging; AA, anterior aortopexy.

Case presentation

We conducted a retrospective review of a prospectively maintained de-identified database of nine consecutive patients with IAS treated by AA from July 2017 to November 2020. The patients were operated on independently by two surgeons at different universities. No comparative approach to an AA was attempted. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patients for the publication of this case series and accompanying images. A copy of the written consent is available for review by the editorial office of this journal. No portions of the manuscript nor the submitted pictures have any information that can lead to the identification of the patient as per the Healthcare Insurance Portability and Accountability Act of 1996. Institutional review board waivers were provided by both the Icahn School of Medicine at Mount Sinai and University of Miami.

Patients were referred by pediatric cardiology or pediatric pulmonology. Initial workup included a detailed history, physical exam, bronchoscopy airway evaluation, and computed tomography (CT) or magnetic resonance imaging

Highlight box

Key findings

- A partial upper median sternotomy provides a versatile approach for an anterior aortopexy (AA) to treat innominate artery compression syndrome (IAS).

What is known and what is new?

- Multiple approaches for an AA have been reported (e.g., left anterior thoracotomy, median transverse incision, full sternotomy) with varying degrees of symptom resolution.
- In this study (N=9) we detail our technique to performing an AA to treat IAS via a partial median sternotomy which resulted in full symptom resolution among 78% (7/9) of patients.

What is the implication, and what should change now?

- We encourage providers to consider a partial upper median sternotomy when planning an AA for IAS.

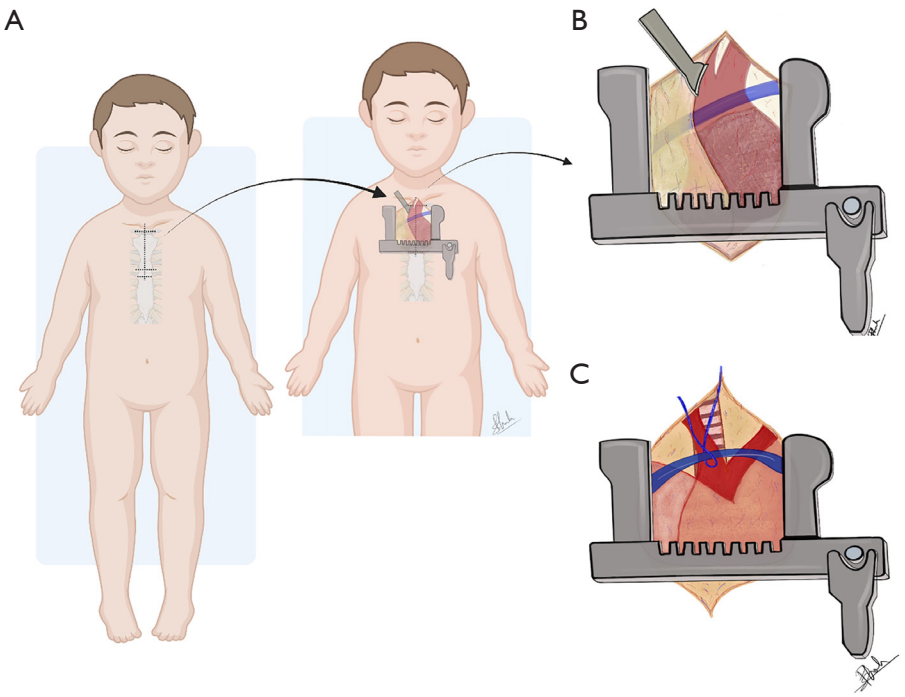

Figure 2 Surgical technique (1/2). (A) Landmarks for an upper partial median sternotomy. The incision is carried through the midline of the sternum generally to the 3rd intercostal space. Critical for exposure is an adequate extension of the neck with a good size shoulder roll, especially in infants. (B) The left lobe of the thymus has been removed, exposing the innominate vein, the innominate artery, the left carotid artery, and the proximal aortic arch covered by the pericardial investment. (C) A full thymectomy is subsequently performed by removing the right lobe and is a key step as it increases the anterosuperior mediastinal space. A limited superior pericardiotomy is performed to expose the junction of the innominate artery and the aortic arch. It is critical to not circumferentially dissect the innominate artery. Image made using BioRender.com and Procreate.

(MRI) (*Figure 1A*). Cases were discussed in multidisciplinary conferences involving cardiac surgery, pediatric cardiology, pediatric pulmonology, and pediatric otolaryngology. Surgery was offered when there was >70% tracheal stenosis at bronchoscopy from anterior tracheal compression by the innominate artery with associated symptoms. CT and MRI imaging were further utilized to assess the anatomy and cause of tracheal compression. Technical success was defined as ≤20% residual stenosis after AA. Symptom continuation or resolution was determined by the surgeon during the last documented clinic visit.

The data were abstracted for demographic information, clinical characteristics, operative intervention, complications, and follow-up (i.e., the last clinic visit in the database). Given the sample size (N=9), only descriptive statistics were used.

Surgical technique

The procedure is performed under general anesthesia with a single lumen endotracheal tube, a radial arterial line for hemodynamic monitoring, and peripheral venous access. A flexible bronchoscopy assesses tracheal compression severity and ensures the endotracheal tube is proximal to the compression (*Figure 1B*). Adequate neck extension with a shoulder roll is critical to facilitate a partial upper sternotomy, especially in infants who often have very short necks. The lower face, neck, and anterior chest is prepped and draped in standard sterile fashion. Prophylactic cefazolin is administered prior to the operation and continued for 24 hours post-operation (9,10).

A partial upper median sternotomy to the third intercostal space is performed using a Diethrich sternal saw. Given the flexibility of the sternum in children, only a central split of the manubrium and the upper sternal body is necessary (*Figure 2A*).

A total thymectomy is then performed. We believe this is critical, as the thymus is often large and acts as a space-occupying lesion. We avoid entering the pleural spaces.

The innominate vein is mobilized and retracted superiorly. The innominate artery is dissected on its anterior aspect and the base of the pericardial reflection on the aortic arch is opened in a limited fashion. It is critical not to circumferentially dissect the innominate artery, as this will lead to a loss of the natural adhesion between it and the anterior tracheal surface. Pulling the innominate artery anteriorly towards the sternum, utilizing the space provided by the thymectomy, relieves the tracheal compression and supports the associated tracheomalacia by suspending the anterior wall of the trachea indirectly to the sternum (*Figure 2B*,*2C*).

Typically, three sets of anchoring sutures are required. The first set is placed at the junction middle and proximal thirds of the innominate artery, the second set is placed at the base of the innominate artery near the aortic arch, and the third set is placed 0.5–1.0 cm below the second set on the anterior surface of the proximal aortic arch. Additional sutures may be placed to optimize the result. Pulling the anterior surface of the innominate artery and the arch with real-time flexible bronchoscopy can guide suture placement.

To ensure a good purchase on the artery, horizontal mattress felt pledgeted sutures with 5-0 Prolene™ sutures (Ethicon, Somerville, NJ, USA) are used on the artery in a partial thickness manner as the anchoring sutures. Using these felt pledgets as anchors, 4-0 or 3-0 Ethibond® excel (Ethicon) suspension sutures are passed through the sternal table to pull the innominate artery anteriorly towards the posterior aspect of the sternal table. If the sternal table is particularly thick, as in an older child, a French eye needle is used to facilitate the suture passage. The composite suture technique enables the use of fine sutures supported with pledgets on the arterial structures as anchoring sutures, and the use of robust sutures with a stronger needle to pass through the sternal table as a suspension suture (*Figure 3A*,*3B*). If the suspension sutures break during the securing process, a new suture can be easily placed. For a left aortic arch, the left half of the sternal table is chosen and vice versa. Before tying down the Ethibond sutures, a mediastinal Blake® drain (Ethicon) is placed to drain the mediastinum. The tip of drain is tucked into the pericardial space and the drain exits below the xiphoid process (*Figure 3C*). Two to three sternal wires are placed, and the sternal tables are partially brought together, leaving a 1 cm gap.

With bronchoscopic guidance, the Ethibond® excel sutures are cinched up and tied on the anterior aspect of the sternal table. Simultaneously, the innominate artery and aortic arch can be observed moving up towards the posterior aspect of the sternal table. The aortic arch suture is tightened first because it is the most robust suture, followed by the sutures distal to it. The innominate artery should rise and be well opposed to the posterior sternal table to optimize the AA. In one case, there was a cut through the anchoring suture on the innominate artery requiring a revision. With all the sutures tightened and tied, the anterior compression on the trachea should be relieved with ≤20% residual tracheal stenosis (*Figure 3B*). If there is >20% residual stenosis, alternate causes of compression need to be examined. These can range from an innominate artery arising further leftward on a left aortic arch, which cannot be addressed by an aortopexy or an intrinsic tracheal abnormality. In such cases, the partial sternotomy is converted to a full sternotomy to evaluate the anatomy further.

The partial sternotomy is closed in a standard fashion with steel wires (*Figure 3C*). The patients are typically extubated on the table and cared for in the intensive care unit (ICU) or step-down unit for the first 24 hours. A portable chest X-ray is obtained to rule out any pneumothorax, assess the drain position, and look for phrenic nerve palsy.

Results

Nine patients underwent AA for IAS (*Table 1*). The median age was 9 months [interquartile range (IQR), 3–16.5 months]. The male to female ratio was 1.25. The aortic arch was left-sided in 89% (8/9) of patients. One patient had Trisomy 21 and another Trisomy 18. All patients had >70% compression by flexible bronchoscopy. Two patients had previous surgeries. One patient had a double aortic arch with an atretic left arch which was divided previously via left thoracotomy. Another patient had a trachea-esophageal fistula which was previously repaired via right thoracotomy. The median follow-up was 6 months (IQR, 4–8.5 months). The surgical indications were: ALTEs 44% (4/9 patients), recurrent intubation 44% (4/9 patients), and severe stridor with >70% luminal reduction 11% (1/9). IAS was a technical success (defined as ≤20% residual stenosis) in 78% of the patients (7/9 patients). Among the seven patients with technical success, 71% experienced complete symptom resolution (5/7). Among the two patients with incomplete symptom resolution despite technical

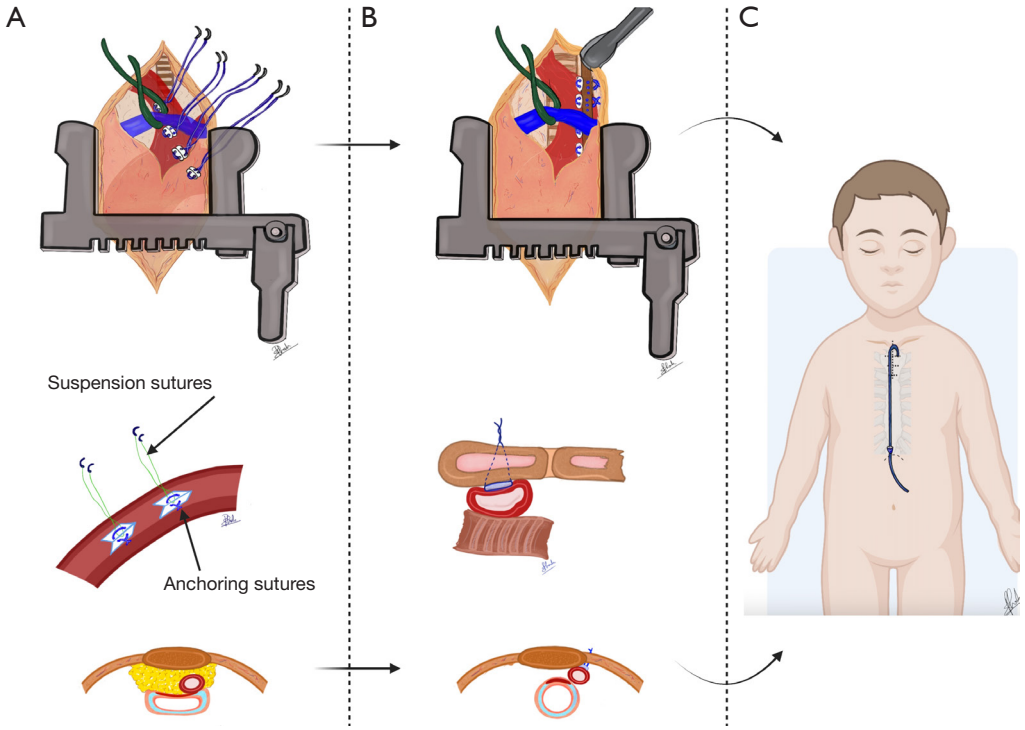

Figure 3 Surgical technique (2/2). (A) The pledgeted anchoring sutures on the innominate artery and proximal aortic arch shown. Using these anchoring sutures, the Ethibond suspension sutures are then placed through them. The inset below shows the compression of the trachea by the innominate artery and the presence of the thymus (yellow structure) acting as a space-occupying structure in a cross-sectional view. (B) The Ethibond suspension sutures have been passed through the left half of the sternal table and tied. Not shown are the sternal wires and the partial approximation of the sternum for clarity. The inset below shows the relief of the tracheal compression, anchoring of the innominate artery to the posterior aspect of the sternum, and absence of the thymus in a cross-sectional view. Sagittal view after aortopexy showing the trans-sternal passage of suspension sutures and relief of the tracheal compression (right figure). (C) Partial sternotomy incision after closure with a Blake drain draining the mediastinal and pericardial space and exiting inferior to the xiphoid process. Image made using BioRender.com and Procreate.

success, one had 20% residual stenosis with mild stridor but no further episodes of ALTE at 6-month follow-up. Mild stridor often improves over time and requires patience and reassurance to family members. The other patient had a 15% residual stenosis with mild residual stridor. This patient also had an associated glottic pathology, potentially contributing to the residual stridor and is awaiting vocal cord surgery.

AA was unsuccessful in two patients with an AA producing 50% residual stenosis. One patient with an unsuccessful AA, had an isolated tracheal cartilage deficiency which was not diagnosed preoperatively. The patient was missing a tracheal ring 0.5 cm above the carina resulting in an entirely membranous section of trachea. The missing ring was not diagnosed preoperatively, as repeat flexible bronchoscopy could easily pass through the stenosis, suggesting an extramural pathology. Furthermore, CT scans taken while the patient was intubated, showed the innominate artery proximal to the anterior trachea (11). The patient was treated with tracheal resection and anastomosis using cardiopulmonary bypass. The other patient with an unsuccessful AA, had an innominate artery arising very leftward along the aortic arch and needed a re-implantation of the innominate artery rightward of the trachea. Interestingly, this patient also had a right-sided upper partial anomalous pulmonary venous return needing a Warden repair. Both patients had complete symptom resolution. There were no wound infections, phrenic nerve palsy, pericardial or pleural effusions. One patient needed a repeat full sternotomy 6 months after the AA to repair a

Table 1 Demographics, clinical presentation, and operative procedure

Patient No.	Age (months); sex	Symptoms	Comorbidities	Aortic sidedness	Severity	Surgical technique	Technical result	Other procedures	Clinical follow-up
1	4; male	ALTE; noisy breathing	None	LAA	75% distal tracheal compression	Thymectomy; anterior aortopexy	10% residual tracheal stenosis	None	Symptom free at 5-month follow-up
2	2; male	Respiratory failure; recurrent intubation	Down syndrome; CAVCD; prune belly	LAA	80% distal tracheal compression	Thymectomy; anterior aortopexy	10% residual tracheal stenosis	Redo sternotomy; CAVCD repair at 8 months of age	Symptom free at 18-month follow-up
3	9; male	ALTE	Trisomy 18	LAA	80% distal tracheal compression	Thymectomy; anterior aortopexy	5% residual tracheal stenosis	None	Symptom free at 3-month follow-up
4	2; female	Recurrent intubation	Prematurity; partial anomalous pulmonary venous return; PDA	LAA	90% distal tracheal compression	Thymectomy; anterior aortopexy	50% tracheal stenosis	Translocation of innominate artery; Warden repair of PAPVR; PDA ligation	Symptom free at 8-month follow-up
5	16; female	ALTE; noisy breathing	None	LAA	80% distal tracheal compression; Mild posterior membrane intrusion	Thymectomy; anterior aortopexy	20% residual tracheal stenosis	None	Some residual stridor, no reflex apnea at 6-month follow-up
6	10; male	ALTE; noisy breathing	None	LAA	80% distal tracheal compression	Thymectomy; anterior aortopexy	5% residual tracheal stenosis	None	Symptom free at 2-month follow-up
7	2; female	Recurrent intubation	Prematurity	LAA	90% distal tracheal stenosis	Thymectomy; anterior aortopexy	50% tracheal stenosis	Full sternotomy; CPB; tracheal resection and anastomosis	Symptom free at 5-month follow-up
8	24; male	Recurrent intubation	Tracheostomy; prior complete vascular ring division; multiple vocal cord surgeries	RAA	80% right lateral distal tracheal compression	Thymectomy; anterior aortopexy; (right sternal table)	15% residual tracheal stenosis	Off ventilator, with tracheostomy	Some residual stridor, awaiting further vocal cord/subglottic surgery for decannulation
9	17; female	Severe stridor	Prematurity; TEF; status post TEF repair via right thoracotomy	LAA	75% distal tracheal stenosis	Thymectomy; anterior aortopexy	10% residual tracheal stenosis	None	Symptom free at 4-month follow-up

ALTE, acute life-threatening event; LAA/RAA, left/right aortic arch; CAVCD, complete atrioventricular canal defect; PDA, patent ductus arteriosus; PAPVR, partial anomalous pulmonary venous return; CPB, cardiopulmonary bypass; TEF, tracheoesophageal fistula.

complete atrioventricular canal defect at 8 months of age.

Discussion

In this paper, we describe our technique for AA selectively for IAS, which resulted in complete symptom resolution in 78% of patients (7/9). This is our preferred approach for the following reasons: firstly, a partial upper median sternotomy facilitates a complete thymectomy. The bilateral phrenic nerves are better visualized and thus at a lesser risk for injury. In our experience, a complete thymectomy is central to the operation's success, as it increases the available anteroposterior dimension in the upper mediastinum. Without a thymectomy, the degree of innominate artery movement towards the sternum and distracting force on the trachea is limited. It is worth noting that successful AAs for IAS have been reported without a thymectomy including via an anterior inframammary incision (12). In addition, Isik *et al.* reported a case series utilizing an upper partial sternotomy and only removing the left part of the thymus gland (13). Secondly, the flexibility of a partial upper median sternotomy increases the chances of a good anatomic result (i.e., ≤20% residual stenosis). In our study, two patients needed alternative procedures. If a ≤20% residual stenosis is not achieved, the surgeon needs to identify alternative factors such as an intrinsic tracheal pathology. Unlike a thoracotomy approach, a partial sternotomy can easily be converted to a full sternotomy enabling the exploration of the anterior surface of the trachea between the superior vena cava and the aorta. In one case, the patient had intrinsic tracheal stenosis due to isolated tracheal cartilage deficiency, which required a tracheal resection (11). Thirdly, the anatomy of the innominate artery is better appreciated by a partial upper median sternotomy. An alternative operation to treat IAS is an innominate artery reimplantation. The compressing innominate artery can be reimplanted in an off-pump manner 1 cm proximal on the greater curvature of the aorta rightward of the trachea (2,3). Such a reimplantation has an immediate effect of relieving the compression with no long-term consequence (2). Only one patient needed this approach in our series after a test AA failed to produce an optimal result. Fourthly, a full sternotomy is unnecessary as the operative procedure is limited to the upper mediastinum.

AA by an upper partial median sternotomy does not preclude a repeat sternotomy if precautions are taken. In an infant with Trisomy 21, prune belly syndrome, complete atrioventricular canal defect, and recurrent failure to wean from mechanical ventilation, an AA was performed at age 2 months and a complete repair of the canal defect was performed at 8 months. The patient did well after the aortopexy and was liberated from mechanical ventilation. At redo sternotomy, because the lower pericardium and the sternum were intact after the initial surgery, an easy retrosternal plane could be established and the posterior sternal table in the region of the aortopexy was divided under direct vision by deviating rightwards of it. The repair was intact and did not interfere with the canal repair. Fixing the innominate artery to the left sternal table in a left-sided aortic arch (and vice-versa in a right-sided aortic arch) helps facilitate a potential redo sternotomy, as the innominate artery is not fixed across the sternotomy incision.

Another critical step for a successful AA is to not develop a paratracheal plane. By dissecting the innominate artery circumferentially away from the trachea, the compressive effect will be relieved. However, if severe tracheomalacia is present, it will lead to the collapse of the anterior wall of the trachea. Pulling the innominate artery anteriorly produces a distracting compression-relieving effect on the trachea and has an additional suspensory effect (1).

The composite suture technique simplifies the operation. While fine sutures are used on the artery, heavy sutures are used for the AA. If the suspension sutures were to snap, they can be easily replaced before the sternum is closed.

The strengths of the study are the description of the technical pearls and pitfalls for a successful AA via an upper partial sternotomy, as evidenced by the good operative outcomes. The limitations are the small sample size, absence of a comparison cohort, and lack of long-term follow-up.

Historically, a left anterior thoracotomy has been the preferred approach utilized in ~70% of AAs for tracheomalacia (7). In an analysis of 100 patients undergoing aortopexy, no differences in outcomes (e.g., mortality, length of ICU stay) was found between a median sternotomy (n=89) and a thoracotomy/thoracoscopic approach (n=11) (14). Wine *et al.* reported a case series including 17 patients with a right median transverse incision. In the study, they reported a complete response in 13 of 21 patients (62%) with the remaining (38%) experiencing a partial response (15). In cases of tracheal compression by the innominate artery at/above the thoracic outlet, a cervical approach can also be utilized (6). When utilizing a suprasternal incision for an aortopexy, Haveliwala *et al.* reported 41% of patients with complete symptom resolution, 32% with improved symptoms, and 27% with no improvement at 6-week

follow-up (N=22) (16). Specifically for IAS, Grimmer *et al.* reported a case series of 22 patients treated with an off-pump innominate artery reimplantation utilizing a median sternotomy approach. Nineteen of the 22 patients (86%) experienced complete symptom resolution, two experienced partial resolution, and one patient experienced no improvement and continued to suffer attacks of severe stridor (2). Our preferred technique, utilizing a partial upper median sternotomy, yielded a 78% technical success rate and 78% rate of complete symptom resolution.

Conclusions

An upper partial sternotomy approach provides a very versatile approach to an AA for IAS. Besides facilitating an adequate AA, it provides options for direct tracheal surgery or an innominate artery reimplantation if an optimal result is not obtained by an AA. We recommend surgeons consider an upper partial sternotomy when performing an AA for IAS.

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Footnote

Reporting Checklist: The authors have completed the AME Case Series reporting checklist. Available at https://tp.amegroups.com/article/view/10.21037/tp-23-597/rc

Peer Review File: Available at https://tp.amegroups.com/article/view/10.21037/tp-23-597/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tp.amegroups.com/article/view/10.21037/tp-23-597/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patients for the publication of this case series and accompanying images. A copy of the written consent is available for review by the editorial office of this journal. No portions of the manuscript nor the submitted pictures have any information that can lead to the identification of the patient as per the Healthcare Insurance Portability and Accountability Act of 1996. Institutional review board waivers were provided by both the Icahn School of Medicine at Mount Sinai and University of Miami.

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