Abstract

Background: Vascular anomalies are rare abnormalities which present with inspiratory stridor and recurrent respiratory tract infection. They are the commonest causes of mortality and morbidity in children due to misdiagnosis. They comprise about less than 4% of congenital heart diseases. The commonest of these anomalies were vascular ring and pulmonary sling.

Objectives: The objective of this study was to determine the prevalence, pattern of presentation and surgical outcomes of children presenting with vascular ring and pulmonary sling.

Methods: A cross-sectional retrospective study in which a review of the records of all children attending Fortis hospital over a 3-year period (2012-2015) was undertaken.

Data were analyzed using SPSS 20. Frequencies, rates and proportions were represented in tables.

Children with confirmed diagnosis of vascular rings and pulmonary slings are included while other causes of stridor were excluded from this study.

Result: A total of 1200 children had open heart surgery in the hospital over a three-year period. Of these, 2 had vascular ring and 3 had pulmonary sling giving a prevalence of 0.4%. Out of the 5 cases, 3 (60%) were male and 2 (40%) female. Male to female ratio was 1.5:1. The mean age of presentation was 2.46±3.52 months. There were three neonates and two infants. All presented with stridor. The mean number of days spent postoperatively was 8±3 days and tracheal stenosis was the only complication noted.

Computerized axial tomography (CT) scan of one of the subjects with vascular ring showed a dilated Kommerells diverticulum (KD).

Conclusion: Vascular ring and pulmonary sling are rare congenital abnormalities seen in our center, early identification and repair will help avert numerous complications that follow it.
pulmonary sling can also give rise to obstructive symptoms such as emphysema and atelectasis [5]. Thus, early recognition and diagnosis of pulmonary artery sling is crucial [6].

Vascular rings encompass only 1–3% of all congenital heart disease [6]. Some vascular rings are associated with other congenital heart lesions while others are isolated defects. Aberrations of tracheobronchial trees are rarely seen with vascular rings but are common in pulmonary artery slings [7].

The diagnosis of vascular rings and pulmonary sling are protean. Barium swallow is the diagnostic procedure of choice. An anterior indentation of the esophagus on the lateral projection is diagnostic of pulmonary artery sling [8]. Furthermore, magnetic resonance imagery or angiography, computerized axial tomography scanning, or a combination can be helpful in delineating the details of the anatomy, as well as 3-dimensional reconstruction of the anatomy of the pulmonary sling as it relates to the airway anatomy [8]. Echocardiography remains the less invasive diagnosis of choice [9–11]. However, examination of the right pulmonary artery reveals the left pulmonary artery arising from its posterior surface.

This retrospective study is crucial so as to enable the pediatrician, cardiologists and cardiac surgeons have a high index of suspicion of its existence (especially in children presenting with stridor at an early age) and to be equipped with the skills to tackle the management and understand the numerous complications that follow the disease. Early identification and appropriate intervention can significantly improve the quality of life.

Prevalent studies of vascular rings and pulmonary slings anomalies are useful to establish baseline rates and to monitor trends over time. They may also help in health services planning and evaluating antenatal screening in populations with high risk. We are not aware of any study of this nature in Fortis hospital, Mumbai. It is hoped that this study will add to the body of knowledge available on these disorders and may stimulate further research in the area on the subject (Figures 1,2).

**Materials and Methods**

The aims of this study were to determine the prevalence of vascular rings and pulmonary slings among children who attended and then admitted in Fortis hospital, Mumbai. It also aims at determining the pattern of presentation, surgical outcomes and follow up states of the subjects.

The study was conducted at the Pediatric cardiac surgery department, Fortis Hospital over a three-year period, from 2012 to 2015. The department of Pediatric cardiac surgery in Fortis is a budding department which is just three years old. Diagnosis was made with the use of Computerized axial tomography scan and Echocardiography.

**Study design**

A cross-sectional retrospective study in which a review of the records of all children attending Fortis hospital over a 3-year period (2012–2015) was undertaken. The diagnosis of pulmonary sling and vascular rings were based on clinical evaluation, echocardiogram and computerized axial tomography. The prevalence rate was estimated as a ratio of the total number of children diagnosed with pulmonary sling and vascular rings to the number of children with open heart surgery in the hospital at duration of study.

Children with confirmed diagnosis of vascular rings and pulmonary slings are included while other causes of stridor were excluded from this study.

**Consent**

Both written and informed consent were obtained from the parents and caregivers before admission, investigations and surgical intervention on these subjects.

**Data analysis**

Data were analyzed using SPSS 20. Frequencies, rates and proportions were represented in tables.

**Results**

A total of 1200 children were operated in the hospital over a three-year period. Of these, three were diagnosed with pulmonary sling and 2 with vascular ring giving a prevalence of
in right pulmonary artery was closed directly with 6-0 prolene. The left pulmonary artery was transected from the right pulmonary artery. Opening was made in the aortic root. On the arrested heart, the left pulmonary artery was dissected along its entire course up to the hilum. Heart was arrested with Saint Thomas cardioplegia cooled to 32°C, and a normal sinus rhythm was maintained.

The patient was put on Cardiopulmonary bypass and the left pulmonary artery origin was identified and visualized, dissected with direct anastomosis done using 7-0 prolene suture. This was divided and ligated while the Ventriculo septal defect (VSD) was also closed.

**Surgical details of Vascular Ring Repair:**

In case 1, the left subclavian artery was well mobilized up to the origin of its branches. Descending aorta was mobilized and looped. Kommerells diverticulum and ligamentum arteriosum dissected and looped. Ligamentum arteriosum ligated with no.2 silk. Cross clamp was placed across the KD at its origin from the descending aorta. Kommerells diverticulum was incised and divided between clamp and Ligamentum arteriosum ligature. Cut end of Kommerells diverticulum was also sutured with 6.0 prolene 10mm suture and reinforced with 6.0 prolene 13mm running suture.

For case 2, For Vascular Ring with aberrant subclavian artery and Right aortic arch, a Left posterolateral thoracotomy through 4th intercostal space was made. The left subclavian artery was well mobilized up to the origin of its branches. Descending aorta was mobilized and looped. Ligamentum arteriosum was dissected and looped. Ligamentum arteriosum ligated with no.2 silk and divided. 'C' Clamp was applied at the base of left subclavian artery and arch, ostia were opened along the anterior surface. Ostial shelf visualized, dissected with direct anastomosis done using 7-0 prolene suture.

**Discussion**

Vascular rings and pulmonary slings are uncommon anomalies that make up less than 1% of all congenital cardiac defects. They occur with about equal frequency in both sexes [10]. This rarity is also confirmed in our setting, having seen five cases in three years with a prevalence also less than 1% (0.4%). Backer [12] et al in his series, described pulmonary artery sling as a rare congenital vascular anomaly and noted only twelve cases in thirty-seven years among infants.

Most of our series presented at neonatal age or early infancy with respiratory symptoms especially stridor. These features were also recorded by Turner [13] et al, though he noted that a minority still presents much later.

Symptoms of airway obstruction are usually seen in patients with this anomaly especially within first few years of life. They include slow breast or bottle feeding, fatigue with feeding, continuous regurgitation, and aspiration pneumonias [14]. Most of our series also presented with poor suck at birth. The respiratory symptoms elicited by these patients could be due to either external tracheal compression or severe tracheal stenosis following complete rings [14]. It is pertinent to note that some vascular rings are associated with congenital heart defects [11]. This is corroborated in this study where 40% of children with pulmonary sling presented with various congenital heart diseases. These include Patent ductus arteriosus (PDA), Ventriculo septal defect (VSD) and Cotriatum sinitrum (CS) all occurred in equal distribution of 20%. Over half (50%) of the subjects with vascular sling had these associated anomalies. Stridor is the commonest symptom (80.0%). The mean age at diagnosis and mean duration of extubation were $5.4 \pm 3.75$ days respectively. Tracheal stenosis was the only complication noted (Table 3).

**Surgical details of pulmonary sling repair**

All the pulmonary slings cases were done on Cardiopulmonary bypass (CPB) through median sternotomy. The subjects were cooled to 32°C, and a normal sinus rhythm was maintained. The left pulmonary artery origin was identified and was seen to arise posteriorly from the right pulmonary artery (RPA). The patient was put on Cardiopulmonary bypass and the left pulmonary artery was dissected along its entire course up to the hilum. Heart was arrested with Saint Thomas cardioplegia given in aortic root. On the arrested heart, the left pulmonary artery was transected from the right pulmonary artery. Opening in right pulmonary artery was closed directly with 6-0 prolene 13mm sutures. The left pulmonary artery which was coursing behind trachea was brought in front of trachea i.e. between trachea and aortic arch. Opening was made in MPA. End to side anastomosis of left pulmonary artery to main pulmonary artery was done using 6-0 10 mm prolene sutures.

In case 1, table 2, in view of cotriatum sinitrum, right atrium was opened, the membrane was visualized and excised.

In case 3, table 2, in view of documented tracheal compression on Computerized axial tomography and perioperative bronchoscopy, aortopecty was done where in after coming off Cardiopulmonary bypass and decannulation, the aortic arch was hitched up on the underside of sternum. Also in case 3, table 2, who had Patent ductus arteriosus (PDA), this was divided and ligated while the Ventriculo septal defect (VSD) was also closed.
Symptomatic newborns and infants with these complex lesions have a high mortality rate without surgical intervention. The ideal operation of pulmonary sling and vascular rings remains debatable, with issues rising from focusing on the need for left pulmonary artery re-implantation and the technique of tracheal reconstruction.

We noted just one case of congenital tracheal stenosis in our study. Congenital tracheal stenosis is a rare condition seen in 30–50% of cases, interestingly, a left pulmonary artery sling may be found with a short segmental stenosis.

Though we noted associated maternal diabetes or hypothyroidism with pulmonary sling, we could not explain if there is any link between these disease and pulmonary sling. This association is also lacking in literatures. [12–14]

There appears a male predominance in both vascular rings and pulmonary slings as seen in our study. Yoon [17] et al also confirmed this gender dominance. The reason for this male predominance could not easily be ascertained, though female double endowment of chromosome X could be suggestive.

It is important to take cognizance of the fact that though echocardiography could be helpful in investigating vascular rings and pulmonary slings. It may not have been able to delineate appropriately if there is any bronchial obstruction or bronchial rings.

This is buttressed by the fact that our case on aberrant subclavian artery was missed with.

### Echocardiography

The use of Computerized axial tomography scan and magnetic resonance imagery remains a gold standard as supported in our findings. This was also corroborated in other studies [16–19].

Definitive managements of vascular rings and pulmonary slings are surgical. Our cases underwent surgery, spent less than two weeks in the hospital after surgery, except one case who spent three weeks because of respiratory obstruction after surgery. However, she was discharged and is presently doing well.

Surgery should be performed promptly after the diagnosis is made, especially in patients with stridor, apnea, or other symptoms of respiratory distress [18]. Delay in operative intervention can result in complications of a serious nature [18].

Left thoracotomy is the surgical approach of choice for the division of a vascular ring in the majority of cases. Anomalous left pulmonary artery has been corrected using the left thoracotomy; however, the use of median sternotomy and cardiopulmonary bypass are known to yield better outcomes [18]. The extremely rare configurations associated with left aortic arch and right descending thoracic aorta are the lesions that should be approached via a right thoracotomy for division of the ring [18].

The major contributor to postoperative mortality is the high frequency of bronchial and tracheal abnormalities in this group of patients [20]. Only one of our patient with pulmonary...
sling had bronchial obstruction and audible stridor which has gradually reduced after treatment.

We noted Kommerell diverticulum (KD) in one of our series. This is a bulb-like out-pouching at the origin of the subclavian artery and ligamentum arteriosum [21]. The major complication of KD is the presence of aneurysm of aorta observed in 3 to 8% of the patients with aberrant subclavian artery [8]. The risk for rupture is variable with incidence of 19 to 53% reported in some series [21].

The importance of bronchoscopy as both a diagnostic modality and a monitor of successful surgical manipulation is vital. As noted in this study, failure to diagnose and/or to treat the pre or intra-operative development of airway narrowing may result in respiratory failure in the very postop and failure to wean the patient from the mechanical ventilation [22,23]. We used bronchoscopy for the aortopexy in order to reduce the need for reoperation and to make intraoperative surgical decision-making easier. Intraoperative bronchoscopy is known to improve the prognosis for favorable clinical outcomes in children with vascular rings and airway obstruction.

The rationale for aortopexy depends on the severity of the condition being treated [22]. In children with severe respiratory failure due to compression of the trachea, aortopexy is the only dependable treatment option.

Conclusion

Vascular ring and pulmonary sling are rare congenital abnormalities seen in our center, early identification and repair will help avert numerous complications that follow it.

Declaration

Ethics approval and consent to participate: This complies with national guidelines [23]. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standard. Fortis research committee and with the institutional guidelines. Fortis research committee and with the ethical standards of the institutional and/or national guidelines [23]. All procedures performed in accordance with national guidelines [23].

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Author contributions

JMC, VA and GS conceived and designed this study while SG and BT helped in diagnosis and, critical revision of the article. JMC also did the Data analysis/interpretation.

References


