Case Report

Management of tracheomalacia in an infant with Tetralogy of Fallot

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ABSTRACT

Most infants with tracheomalacia do not need specific therapy as it usually resolves spontaneously by the age of 1–2 years. Severe forms of tracheomalacia characterized by recurrent respiratory infections require active treatment which includes chest physiotherapy, long term intubation or tracheostomy. Aortopexy seems to be the treatment of choice for secondary and even primary forms of severe tracheomalacia. Itentails tracking and suturing the anterior wall of the aorta to the posterior surface of the sternum. Consequently, the anterior wall of the trachea is also pulled forward preventing its collapse. A 3-month-old girl baby who was on ventilatory support for 2 months due to severe tracheomalacia associated with a cyanotic congenital heart disease underwent intracardiac repair and aortopexy along with Lecompte's procedure as all the conservative measures to wean off the ventilator failed. The baby was extubated on the third post-operative day and the post-operative period was uneventful.

Key words: Aortopexy, tetralogy of fallot, tracheomalacia

INTRODUCTION

Tracheomalacia is a structural abnormality of the tracheal cartilage causing collapse of its walls and airway obstruction. Tracheomalacia can be associated with various congenital anomalies, including cardiovascular defects, tracheoesophageal fistula and growth or developmental abnormalities. Primary tracheomalacia is a rare congenital condition characterized by weakness of tracheobronchial cartilaginous bridges, resulting in reduced tracheobronchial lumen.[1] It can be an isolated finding in healthy infants, but it is more commonly seen in premature infants.[2] It is believed to be a consequence of the inadequate maturity of tracheobronchial cartilage, either from premature delivery or from an innate immaturity despite normal gestation which is also the primary form of of tracheomalacia, where in the aetiology is unknown. Secondary forms of tracheomalacia are characterized by other contributing factors like oesophageal atresia, prolonged positive pressure ventilation, tracheostomy, large neck swellings, vascular ring compressions and connective tissue disorders. Primary forms of tracheomalacia very rarely require surgical intervention. Some authors have reported no gender predominance in the primary form of the disease, whereas others have reported a definite male predominance.[3] The following case report is about a 3 months baby who was suffering from a cyanotic congenital heart disease superimposed by tracheomalacia which caused the multiple episodes of cyanosis and desaturation. This baby underwent intracardiac repair along the Lecompte's maneuver and aortopexy.

CASE REPORT

A 3-month-old female baby weighing 5.9 kg was referred to the paediatric intensive care unit for tracheomalacia. Baby was intubated at 20 days of age for respiratory distress and cyanosis. Multiple trials to extubate the child failed and she became ventilator dependent. The baby was ventilated with synchronized intermittent mandatory ventilation (SIMV) pressure

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support mode with peak inspiratory pressure 30 cm of H_2O. Her saturation was 90–92% with FiO2 of 0.5, blood pressure and heart rate were within normal limits. The baby had multiple episodes of desaturation which was treated with positive pressure ventilation using a manual resuscitation bag. Echocardiogram revealed a congenital cyanotic heart disease with a perimembranous ventricular septal defect, moderate infundibular pulmonary stenosis and aortic override of 40%. Computed tomographic scan of the thorax showed consolidation changes involving the apical segments of right upper lobe, lateral segment of right middle lobe, apical and posterior basal segments of right lower lobe. Dynamic reconstruction of the computed tomographic scan showed tracheal narrowing just above the level of the carina [Figure 1]. The child was posted for intracardiac repair with tracheal reconstruction with aortopexy.

Baseline monitors included electrocardiogram, pulse oximetry, invasive arterial pressure and central venous pressure. The child was induced with injection ketamine 15 mg IV and glycopyrrolate 60 μg IV. Adequacy of ventilation was checked and then injection vecuronium 0.5 mg IV for muscle relaxation was administered. The endotracheal tube was changed from oral to nasal (4.5 mm) for better tube tolerance during the post-operative period. Anaesthesia was maintained with oxygen with FiO_2 of 0.50, air, sevoflurane with end tidal concentration of 1–2%, injection fentanyl 5 μg and midazolam at 20 mcg/kg boluses as and when required. Cardiopulmonary bypass was achieved after adequate heparinisation. Huge patent ductus arteriosus was found and was ligated [Figure 2]. Right pulmonary artery was enlarged and compressing trachea. Ventricular septal defect was closed and right ventricular outflow obstruction was relieved. Lecompte's manoeuvre (bringing the pulmonary artery anterior to aorta) was performed to relieve the tracheal compression and this was further stabilized with aortopexy. The patient was weaned off cardiopulmonary bypass with IV infusions of nitroglycerine at 0.5 μg/kg/min and dobutamine at 5 μg/kg/min. The baby was shifted to intensive care unit. She was weaned and extubated on third post-operative day.

DISCUSSION

Tracheomalacia is a process characterized by flaccidity of the supporting tracheal cartilage, widening of the posterior membranous wall, and reduced anterior-posterior airway caliber. These factors cause tracheal collapse, especially during times of increased airflow, such as coughing, crying, or feeding.[4-6] Congenital tracheomalacia are rare and no definite incidence rates are available.[7] The incidence of tracheomalacia in all age group was estimated to be atleast 1 in 2100.[8] Infants present with expiratory stridor which may worsen with supine position, crying, and respiratory infections. Feeding difficulties, hoarseness,

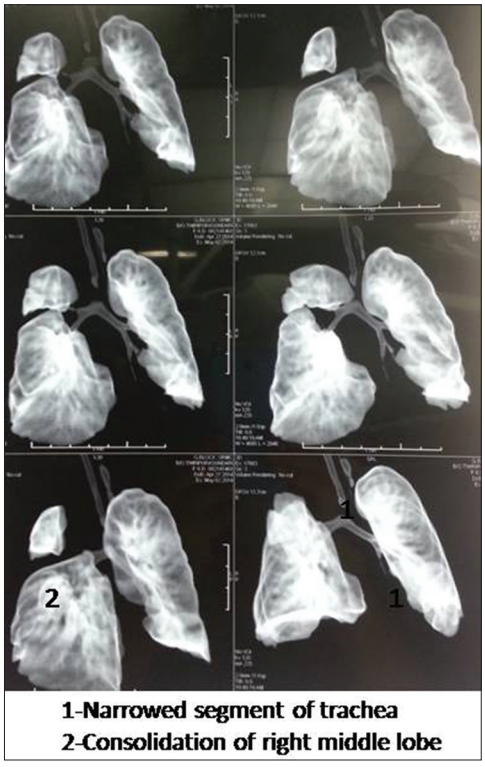

Figure 1: Dynamic computed tomographic scan showing narrowed trachea and consolidation

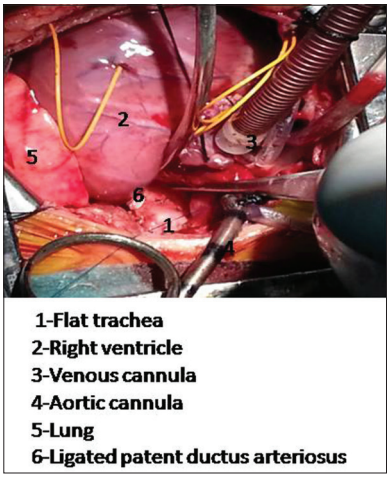

Figure 2: Intraoperative diagram showing ligated patent ductus arteriosus and flat trachea

aphonia, and breathing difficulty; our patient presented with all of the above signs and symptoms.

Tracheomalacia is diagnosed by rigid bronchoscopy in spontaneously breathing patients, chest radiographs and cinefluoroscopy using a contrast through the oesophagus. Dynamic airway collapse is better identified by computed tomographic scans. They are better predictors of tracheomalacia compared to the end expiratory films of chest radiograms. Diagnosis of tracheomalacia in the present case was confirmed using a dynamic computed tomographic film. Generally benign, self-resolving with age and by maturation of airway structures, conservative management is given in the form of chest physiotherapy, humidified oxygen and treatment of the underlying infection. With conservative measures, the symptoms often resolve spontaneously by age 18–24 months.[5] Surgery is only recommended for severe symptoms and failure of conservative therapy.[9] Our patient was kept on SIMV pressure support mode with peak inspiratory pressures of 30 cm of H2O. This did not improve the baby's respiratory condition but further worsened the hypoxia and right to left cardiac shunt which resulted in the multiple episodes of cyanosis as well. Hence, she was taken up for surgery as the final treatment of choice. Surgery includes correction of the underlying cause, such as vascular ring when present, tracheostomy, intraluminal or extraluminal stenting and aortopexy.[10] Aortopexy has been proven to be a safe and effective way to relieve the tracheal compression by increasing the anterior posterior diameter to maintain a patent lumen. Right and left anterior as well as lateral thoracotomy incisions have been used and also cervical, mediastinal and thoracoscopic approaches have been reported. The many techniques indicate that, so far, no one approach has proven superior for all cases. Thoracoscopic aortopexy can be safely performed even in small infants as long as equipment and skill for paediatric thoracoscopy are available. The authors believe that thoracoscopic exposure offers advantages over open technique and is cosmetically more pleasing to patients and their families. It may result in less pulmonary complications, shorter hospital stay, and less narcotic requirement compared to open thoracotomy. But this approach was not preferred for our patient as an intracardiac repair was also necessary for the congenital cyanotic heart disease.

A recent report of aortopexy in 28 children with severe and localized tracheomalacia utilized a left lateral muscle-sparing approach. The indications included acute life-threatening events, failure to extubate and recurrent pneumonia. Most symptoms of tracheomalacia resolved after aortopexy.[11] Lecompte's surgical manoeuvre is used for the correction of transposition of great arteries associated with ventricular septal defect and pulmonary stenosis, where the distal aortic segment is transfixed beneath the bifurcation of the pulmonary artery. This procedure was utilized in our case to relieve the pressure caused by pulmonary artery compression. The Lecompte operation allows complete repair in infancy and may reduce the need for reoperation and the prevalence of residual pulmonary outflow tract obstruction.

CONCLUSION

Presence of cyanotic congenital heart disease and tracheomalacia posed an amalgam of cardiovascular and respiratory problems leading to prolonged ventilation, which started immediately after birth. The right to left shunt in the ventricular septal defect worsened the cyanosis , this was associated with a right ventricular outflow tract obstruction. The presence of tracheomalacia and the hypoxia that resulted from it added on to the underlying cardiovascular disease. Hence, the baby underwent an intracardiac repair along with aortopexy which corrected the primary cause for tracheomalacia.

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REFERENCES

1. Kumar S, Gupta R, Wadhawan S. Anesthetic management of an infant for aortopexy. J Anaesthesiol Clin Pharmacol 2013;29:252-4.
2. Jacobs IN, Wetmore RF, Tom LW, Handler SD, Potsic WP. Tracheobronchomalacia in children. Arch Otolaryngol Head Neck Surg 1994;120:154-8.
3. Adler SC, Isaacson G, Balsara RK. Innominate artery compression of the trachea: Diagnosis and treatment by anterior suspension. A 25-year experience. Ann Otol Rhinol Laryngol 1995;104:924-7.
4. Beasley SW, Qi BQ. Understanding tracheomalacia. J Paediatr Child Health 1998;34:209-10.
5. Carden KA, Boiselle PM, Waltz DA, Ernst A. Tracheomalacia and tracheobronchomalacia in children and adults: An in-depth review. Chest 2005;127:984-1005.
6. Gaissert HA, Burns J. The compromised airway: Tumors, strictures, and tracheomalacia. Surg Clin North Am 2010;90:1065-89.
7. Jaquiss RD. Management of pediatric tracheal stenosis

and tracheomalacia. Semin Thorac Cardiovasc Surg 2004;16:220-4.
8. Boogaard R, Huijsmans SH, Pijnenburg MW, Tiddens HA, de Jongste JC, Merkus PJ. Tracheomalacia and bronchomalacia in children: Incidence and patient characteristics. Chest 2005;128:3391-7.
9. Antón-Pacheco JL, García-Hernández G, Villafruela MA. The management of tracheobronchial obstruction in children. Minerva Pediatr 2009;61:39-52.
10. Kikuchi S, Kashino R, Hirama T, Kobayashi H, Abe T. Successful treatment of tracheomalacia associated with esophageal atresia without a tracheoesophageal fistula by aortopexy: Report of a case. Surg Today 1999;29:344-6.
11. Dave S, Currie BG. The role of aortopexy in severe tracheomalacia. J Pediatr Surg 2006;41:533-7.

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Announcement

CALENDAR OF EVENTS OF ISA - 2015

Certain important dates are given here for the members. All the applications should be sent by registered post (with Acknowledgement Due)

Date	Name of the Award/Post	Application has to be sent to
30th June 2015	Bhopal Award for Academic Excellence	Hony. Secretary, ISA
15th August 2015	Prof. A. P. Singhal Life Time Achievement Award	Hony. Secretary, ISA
31st October 2015	Dr. (Mrs.) Rukmini Pandit Award - Publication format along with Conference Presentation Certificate	Hony. Secretary, ISA
31st October 2015	Y. G. Bhoj Raj Award - Best Review Article in IJA	Hony. Secretary, ISA
31st October 2015	Dr. Kop's Award	Chairman Scientific committee of ISACON with a copy to Hony Secretary ISA
27th November 2015	Dr. TN Jha Memorial & Dr. KP Chansoriya Travel grant	Hony. Secretary, ISA
27th November 2015	Late Dr. Venkata Rao Memorial Oration	Hony. Secretary, ISA
27th November 2015	Ish Narani Best Poster Award	Chairman Scientific Committee ISACON
28th November 2015	ISA GOLDCON QUIZ Competition	Chairman Scientific Committee ISACON
28th November 2015	Awards for 1. Best City Branch 2. Best State Branch 3. Best Metro Branch 4. Public Awarness Individual 5. Public Awarness City 6. Public Awarness State 7. Ether Day State 8. Ether Day City 9. Membership Drive % (State) 10. Membership Drive No.s (State) 11. Individual Drive	Hony. Secretary, ISA

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