RESEARCH PAPER

Clinical Characteristics and Associated Congenital Lesions with Tracheomalacia in Infants

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Objective: To describe clinical presentation, bronchoscopy findings and associated anomalies in cases of congenital tracheomalacia in infants (age <1y). **Methods:** Hospital record review of 88 infants (mean age 8 mo, 57 males) diagnosed as having tracheomalacia by flexible bronchoscopy between 2012 and 2015. **Results:** The predominant features were wheeze (57.9%), stridor (42.1%), cough (38.6%), pneumonia (29.5%) and collapse (12.5%). On bronchoscopy, malacia was observed in lower half of trachea in 51 (57.9%) infants. Synchronized airway lesions observed were laryngomalacia (30.7%) and bronchomalacia (3.4%). 15 (17%) infants had associated congenital heart disease and 21 (23.8%) required care in intensive care unit. **Conclusion:** Wheeze,stridor and cough are the main symptoms in tracheomalacia. Laryngomalacia and congenital heart diseases are the most common other anomalies associated in these infants.

Keywords: *Congenital heart disease, Bronchomalacia, Flexible bronchoscopy,Outcome*.

Airway malacia is a structural anomaly that predisposes children to recurrent respiratory problems. Congenital airway malacias include tracheomalacia, bronchomalacia, and tracheobronchomalacia [1]. Tracheomalacia is due to the developmental defect in the cartilage of the tracheal wall. Since the walls of the trachea are softer than normal, partial collapse of trachea results during expiration, and gets exaggerated during activity [2]. Depending on the site affected, tracheomalacia may present with a wide spectrum of clinical problems from noisy breathing to acute severe wheeze. Children present with respiratory compromise, and require intensive care and ventilator support. Morbidity increases if tracheomalacia is associated with other anomalies. This study was undertaken to describe the clinical presentation, characteristics and associated anomalies of infants with tracheomalacia.

Methods

Records of infants who were diagnosed as congenital tracheomalacia based on the flexible bronchoscopy between 2012 and 2015 at Kanchi Kamakoti CHILDS Trust Hospital, were reviewed. All the bronchoscopy procedures were video recorded with mutual verification by a fellow doctor trained in pediatric pulmonology. Except in two infants, all the bronchoscopy procedures were done under local anesthesia [3]. The observation of the anterior tracheal wall collapsing and reducing >50% of lumen diameter against the posterior wall was the basis of diagnosis of tracheomalacia. In addition to the bronchoscopic details, the spectrum of clinical presentation, associated radiographic and echocardiogram findings were recorded.

Results

Of the 88 infants (mean age 8 mo, 57 boys) with tracheomalacia diagnosed during the study period, 16 (18.2%) were born preterm, out of which five had received surfactant and ventilation for respiratory distress syndrome. Nineteen (21.6%) of infants were aged below 3 months, 20 (22.7%) were in the age group 4-6 months, 16 (18.2%) were aged between 7 and 9 months, and 33 (37.5%) were aged 10 months and above. The predominant complaints were wheeze in 51 (57.9%), stridor in 37 (42.1), and cough in 34 (38.6%) infants. All infants with tracheomalacia had recurrent respiratory problems requiring repeated nebulizations. Radiographic features included pneumonia (consolidation) in 26 (29.5%), collapse in 11 (12.5%), mild cardiomegaly in 2 (2.3%), hypoplasia of lung in 1 (1.2%); 48 (54.5%) had a normal chest *X*-ray. Bronchoscopy demonstrated malacia of the upper half of trachea in 37 (42.1%) and lower half in 51 (57.9%). Synchronized airway lesions were observed in 30 (34.1%) infants, which included laryngomalacia in 27 (30.7%) and bronchomalacia in 3 (3.4%). Congenital heart diseases were noted in 15 (17.0 %), which included atrial septal defect (4), ventricular septal defect (3), patent

WHAT THIS STUDY ADDS?

- A high proportion of hospitalized infants with tracheomalacia have an associated cardiac anomaly.

ductus artoriodus (3) pulmonary hypertension (2), partial anomalous pulmonary venous connection (1), single ventricle (1) and Fallot's tetralogy (1). Twenty-one (23.8%) of infants with tracheomalacia required PICU care.

DISCUSSION

In this hospital record review, we documented that synchronized airway lesions and congenital heart diseases are found commonly in infants with tracheomalacia. The actual incidence of tracheomalacia cannot be arrived at as it was a hospital-based study. Earlier studies report on estimated incidence of at least 1 in 2,100 children [4]. Though some centers use general anesthesia and rigid bronchoscopes, we used flexible bronchoscopy under topical anesthesia. As tracheomalacia is a dynamic condition, flexible bronchoscopy performed under topical anesthesia is the preferred technique [5].

A recent study in infants with moderate and severe laryngomalacia, flexible bronchoscopy demonstrated synchronized lower airway anomalies in 48%, of which tracheomalacia was the most common [6]. Parenchymal lung lesions are also associated with tracheomalacia, and combined therapy to control pulmonary infection along with airway clearance (remove the backlog of secretions) may improve the outcome [7]. A high proportion of congenital heart disease in children with tracheonalacia has also been reported earlier [8-10], and it may be worthwhile screening all such children with echocardiography.

In our series, about one-fourth infants required ICU care for severe wheeze. In airway malacias, due to the defective cartilage, the contour of airways is maintained by the bronchial smooth muscle tone. When these children are treated with beta-agonists, their wheeze may worsen due to a further reduction in the muscle tone [11]. Thus any infant presenting with acute wheeze requiring a hospitalization whose response to treatment is inadequate or worsening with standard protocol warrants bronchoscopy to rule out underlying congenital airway anomalies.

The main limitations of this study are: retrospective data, referral bias, small sample size, and lack of follow-up data and surgical outcomes.

We conclude that infants with tracheomalacia have a high frequency of associated anomalies of airway and cardiovascular system.

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REFERENCES

1. Holinger LD. Etiology of stridor in the neonate, infant and child. Ann Otol Rhinol Laryngol. 1980;89:397-400.
2. Goyal V, Masters IB, Chang AB. Interventions for primary (intrinsic) tracheomalacia in children. Cochrane Database SystRev.2012; CD005304.
3. Austin J, Ali T. Tracheomalacia and bronchomalacia in children: Pathophysiology, assessment, treatment and anaesthesia management. Paediatr Anaesth. 2003;13:3-11.
4 Boogaard R1, 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.
5. Godfrey S, Avital A, Maayan C,Rotschild M, Springer C. Yield from flexible bronchoscopy in children. Pediatr Pulmonol. 1997;23:261-9.
6. Vijayasekaran D, Gowrishankar NC, Kalpana S, Vivekanandan VE, Balakrishnan MS, Suresh S. Lower airway anomalies in infants with laryngomalacia. Indian J Pediatr. 2010;77:403-6.
7. Pan W, Peng D, Luo J, Liu E, Luo Z, Dai J, *et al*. Clinical features of airway malacia in children: a retrospective analysis of 459 patients. Int J Clin Exp Med. 2014;7:3005-12.
8. Masters IB, Chang AB, Patterson L, Wainwright C, Buntain H, Dean BW, *et al*. Series of laryngomalacia, tracheomalacia, and bronchomalacia disorders and their associations with other conditions in children. Pediatr Pulmonol. 2002;34:189-95.
9. Lee SL, Cheung YF, Leung MP, Nq YK, Tsoi NS. Airway obstruction in children with congenital heart disease: Assessment by flexible bronchoscopy. Pediatr Pulmonol. 2002;34:304-11.
10. Chen Q, Langton-Hewer S, Marriage S, Hayes A, Caputo M, Pawade A, *et al*. Influence of tracheobronchomalacia on outcome of surgery in children with congenital heart disease and its management. Ann Thorac Surg. 2009; 88:1970-4.
11. Vijayasekaran D, Kalpana S, Vivekanandan VE. Tracheobronchomalacia presenting as an infantile wheeze. Indian Pediatr. 2011;48:903-4.