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Original Article

The role of flexible fibreoptic bronchoscopy in evaluation of pulmonary diseases in children with congenital cardiac disease

Ori Efrati,[1] Udi Gonik,[1] Dalit Modan-Moses,[1] Bella Bielorai,[2] Asher Barak,[1] Amir Vardi,[3] Gideon Paret,[3] David Mishaly,[4] Amos Toren[2]

[1]Pediatric Pulmonology Unit, [2]Department of Pediatric-Hemato-Oncology and BMT, [3]Department of Pediatric Intensive Care, [4]Pediatric Cardiac Surgery Unit, Safra Children's Hospital, The Sheba Medical Center, Tel-Hashomer, affiliated to the Tel-Aviv University, Sackler School of Medicine, Tel-Aviv, Israel

Abstract *Background:* Diseases of the lungs and airways can be life-threatening in patients with congenital cardiac disease. External compression, and tracheobronchomalacia, as well as pathology of the large airways, can cause respiratory symptoms such as wheezing, stridor, fever, and dyspnoea, along with producing difficulty to wean the patient from mechanical ventilation. A definite diagnosis may be achieved by fibreoptic flexible bronchoscopy. *Aims:* To characterize the patients who underwent bronchoscopy for respiratory diseases associated with congenital cardiac disease, and to evaluate the safety, total diagnostic rate, and diagnostic rate, which yielded alteration of treatment in this group of patients. *Methods:* We reviewed retrospectively the records of 39 patients with congenital cardiac disease associated with abnormalities of the airways who underwent 49 fibreoptic bronchoscopies between 1999 and 2004. The patients were evaluated with regard to rate of diagnosis, the rate of procedures that resulted in alteration of management, and the rate of complications. *Results:* Fibreoptic flexible bronchoscopy was safe, and contributed to a specific diagnosis in almost nine-tenths of the procedures. A specific diagnosis that resulted in treatment alteration was achieved in one-fifth of the procedures. *Conclusions:* Fibreoptic flexible bronchoscopy has an important role in the evaluation of pulmonary abnormalities in patients with congenital cardiac diseases, and should be considered as an initial diagnostic tool that may guide the clinician with regard to the proper therapy.

Keywords: Bronchoscopy; cardiac disease; bronchoalveolar lavage; left main bronchus compression; pneumonia; atelectasis

RESPIRATORY DISEASES, AND IN PARTICULAR obstruction of the trachea and major airways, may be a life-threatening complication in children with congenital cardiac disease. Hence, a quick and accurate diagnosis is needed, so that appropriate treatment can be initiated.[1–4] These abnormalities include pulmonary atelectasis, pulmonary hyperinflation, retained secretions, hypersecretions, abnormalities in gas exchange, and pneumonia.[5–7] Vascular rings, and distended pulmonary arteries, in particular in patients with left-to-right shunts and dilated cardiac ventricles, are the main structural abnormalities responsible for the abnormalities.[2,8–10] Fibreoptic bronchoscopy is an important and safe technique in the evaluation of abnormalities of the lungs and airways, allowing an excellent assessment of dynamics and anatomy.[11–13] The goals of our study, therefore, were to characterize the patients who underwent bronchoscopy for respiratory diseases associated with congenital cardiac disease, and to evaluate the safety, total diagnostic rate, and the percentage of procedures which dictated alterations in treatment for this group of patients.

Correspondence to: Ori Efrati, MD, Pediatric Pulmonology Unit, Chaim Sheba Medical Center, Tel-Hashomer 52621, Israel. Tel: +972 3 5302884; Cell: +526667337; Fax: +972 3 5345914; E-mail: ori.efrati@sheba.health.gov.il

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Patients and methods

We studied retrospectively the records of 39 children with congenital cardiac disease and pulmonary diseases, who underwent 49 fibreoptic bronchoscopies

Table 1. Details of children undergoing bronchoscopy according to cardiac disorders.

Group	TOF	TGA	ABSENT PV	VSD/ASD	AORTIC COARCT	AVSD	PULMON SLING	IHSS	HEART TRPLT.	CAT	TOTAL
No. of patients	11	4	2	6	5	5	1	1	1	3	39
Boys/girls	8:3	2:2	2:0	3:3	4:1	2:3	1:0	1:0	0:1	0:3	23:16
Median age (yr)	4.3	<1	<1	2.3	<1	<1	<1	15	5	6.7	2.7
No. of procedures	14	4	3	10	5	6	2	1	1	3	49

Abbreviations: TOF: tetralogy of Fallot; TGA: transposition of the great arteries; IHSS: idiopathic hypertrophic sub-aortic stenosis; AVSD: atrioventricular septal defect with common atriuoventricular junction; CAT: common arterial trunk; PV: pulmonary valve; VSD/ASD: ventricular and atrial septal defect; Pulmon: pulmonary sling; TRPLT: transplantation

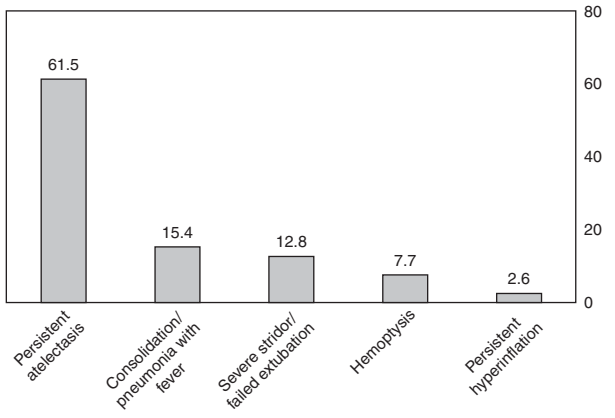

Figure 1.
Indications for fibreoptic bronchoscopy%.

during the period 1999 through 2004. In 10 patients, there were 2 procedures, due to recurrent haemoptysis in 1, persistent fever in 3, atelectasis in 4, and obstruction of the upper airways and failure to extubate in 2. Data were obtained by reviewing the charts and the reports of the bronchoscopies. The cardiac anomalies were diagnosed and evaluated by transthoracic echocardiography with or without angiography (Table 1). All the children were hospitalized in the paediatric intensive care unit before or after cardiac surgical repair, and all had severe respiratory symptoms.

Indications for fibreoptic bronchoscopy (Figure 1)

The indications for bronchoscopy were persistent atelectasis in 24 patients (61.5%), consolidation and pneumonia with fever in 6 patients (15.4%), haemoptysis in 3 patients (7.7%), severe stridor or failed extubation in 5 patients (12.8%), and persistent hyperinflation in 1 patient (2.6%). The airway was studied for intrinsic or intraluminal pathologies, extrinsic compression or pulsation on the airways, and the demonstration of tracheobronchomalacia.

Fibreoptic bronchoscopy

All patients were evaluated by a paediatric pulmonologist, and informed consent was obtained from the parents. All the procedures were performed in the paediatric intensive care unit. Fibreoptic bronchoscopy was performed with a paediatric bronchoscope (Olympus BF Ultrathin 2.2, BF XP40 and BF 3C40, with outer diameter of 2.2, 2.8 and 3.6 mm, Two Corporate Drive, Melville, New York) and all findings were recorded. The patients were anaesthetized with intravenous propofol, or sedated with intravenous midazolam. Topical anaesthesia with 1% lidocaine was applied to the nose, vocal cords and trachea to the patients who were not intubated. The trans-nasal approach was used in all patients who were not intubated, while the endotracheal approach was used in mechanically ventilated patients.

Continuous oxygen, at 3 liters per minute, was delivered via a nasal canula, or 100% oxygen was administrated through the endotracheal tube when the child was mechanically ventilated. Heart rate, blood pressure, and saturations of oxygen were monitored continuously during the procedure. An experienced assistant, usually an attending physician in paediatric intensive care, and a nurse were present during the procedure to evaluate the state of the patient, and to administer medications.

Classification of the results of bronchoscopy

We classified the findings into:

- Bronchoscopy providing a specific diagnosis.
- Bronchoscopy providing a specific diagnosis which resulted in alteration of management, also described as clinical yield.
- Non-diagnostic fibreoptic bronchoscopy.

Results

We performed a total of 49 bronchoscopies in 39 children with congenital cardiac disease and pulmonary abnormalities during the period of the study. Of these, 23 (59%) were male, and the mean age of the overall group was 2.9 plus or minus 4.8 years, with a range from 4 weeks to 19 years. Their congenital cardiac diseases are summarized in Table 1. All

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bronchoscopic procedures followed cardiac surgical repair. Anomalies involving the airways were suspected following chest X-rays, computerized tomography scans, or a clinical picture of stridor or failure of extubation.

Bronchoscopic findings (Tables 2 & 3)

The overall diagnostic rate following fibreoptic bronchoscopy in our patients was 89.8%. Extrinsic compression and obstruction of the airways was the most common cause of anomalies involving the lower airways. We found extrinsic pulsation and compression of the airways in 14 patients (35.9%) (Table 2). The left main bronchus was the most commonly compressed region, seen in 9 of the 14 patients (Table 3). The cardiac and vascular components that caused airway compression were dilated pulmonary arteries, an enlarged left atrium, a dilated aorta, and the pulmonary sling. Infection of the lower airways was diagnosed in 7 cases (17.9%). From these patients, Staphylococcus aureus, Neisseria lactamica, and Streptococcus pneumonia were each isolated in individual cases by bronchoalveolar lavage. Macroscopic and microscopic changes in bronchoalveolar lavage consistent with infection were seen in the 4 additional patients, without the isolation of microorganism.[14,15] Intrinsic instability of the walls of the airways was diagnosed in 6 patients (15.4%), due to tracheomalacia and bronchomalacia in 3 patients each (Table 2). Haemorrhage was detected and lavaged in 2 patients (5.1%), both with tetralogy of Fallot. In 5 patients, we found masses involving the airways, or other narrowings. Bronchial casts occurred in 3, while 2 demonstrated narrowing of the airways subsequent to surgery (Table 2).

Altogether, fibreoptic bronchoscopy yielded a specific diagnosis in 8 patients (20.5%), resulting in alterations in treatment that that led to resolution of the problem. Specifically, 3 patients were treated with antibiotics for bacterial infection, 3 had surgical correction, and 2 had their atelectasis opened (Fig. 2).

In 4 of the 39 patients (10.2%), the procedures yielded no diagnostic utility.

Complications

Complications were recorded during and in the post procedural period, and occurred in 6 of the 49 procedures (12.2%). In 5 patients, the complications were minor including mild desaturation in 4 patients, and mild bleeding in 1 patient, all resolved by supplemental oxygen and lavage of the airway with 0.9% salt solution. Only 1 patient developed severe hypoxemia and bronchospasm requiring cardiopulmonary resuscitation and mechanical ventilation following the procedure. None of the patients died during or immediately after the procedure.

Table 2. Diagnosis by fibreoptic bronchoscopy.

Diagnosis	No. of patients
External compression of airways	14 (35.9%)
Infection	7 (17.9%)
Tracheobronchomalacia	6 (15.4%)
Airway mass/narrowing	5 (12.8%)
bronchial casts	3
narrowing of the airways post surgery	2
Haemorrhage	2 (5.1%)
Pulmonary sling	1 (2.6%)
No diagnosis	4 (10.3%)

Table 3. Diagnosis and site of external airway compression or obstruction.

Site of external airway compression	No. of patients	%
1. Left main bronchus	9	23.1
Atrioventricular septal defect	2	
Ventricular septal defect	2	
Common arterial trunk	1	
Tetralogy with "absence" of the pulmonary valve	1	
Transposition	1	
coarctation of aorta	1	
pulmonary sling and Tetralogy	1	
2. Extrinsic compression of the carina	2	5.1
3. Pulsation of the left lower bronchus	2	5.1
4. Extrinsic compression of the trachea	1	2.6
Total	14	35.9

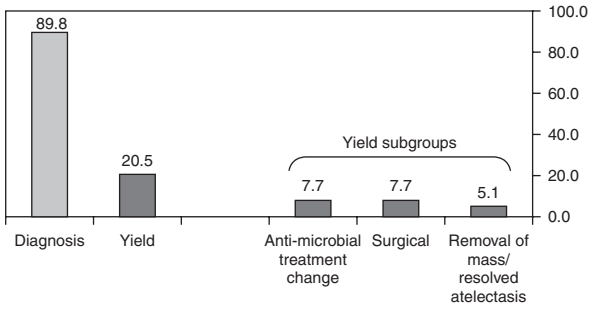

Figure 2.
Diagnosis and yield of fibreoptic bronchoscopy%.

Discussion

Compression and obstruction of the airways, and failure to wean the patient from mechanical ventilation following a cardiac surgery, are relatively common. Furthermore, rates of morbidity and mortality may be high when respiratory distress develops due to compression of the airways.[2,16] As did Lee et al.,[1] we have

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demonstrated that fibreoptic bronchoscopy is clinically useful, and provides, in our hands, a specific diagnosis in nine-tenths of patients with anomalies of the airways associated with congenital cardiac disease. A specific diagnosis with a clinical yield was achieved in one-fifth of the patients, in whom prompt initiation of the indicated treatment resolved the pathology of the airways (Fig. 2). The spectrum of disease observed in our patients is similar to that described by other investigators.[1,2,8,17] Extrinsic compression or pulsation of the airway was the most common cause of anomalies, occurring in just over one-third of the patients. Compression may develop in cyanotic or acyanotic congenital cardiac disease, especially when there is left-to-right shunting.[17] Compression and obstruction of the airways is due to the close relationship between the tracheobronchial tree and the anomalous enlarged heart and vascular structures. Our results, similarly to other studies, demonstrate a high prevalence of vascular compression of the left main bronchus.[1,10,17,18] The left main bronchus passes under the aortic arch, having an intimate anatomic relationship with the pulmonary artery and left atrium. Thus, dilation of the pulmonary arteries due to cardiac abnormality with high left-to-right shunting, as occurs in ventricular septal defect, atrioventricular septal defect, or tetralogy of Fallot with rudimentary formation of the pulmonary valve, may result in compression of the left main bronchus. Moreover, dilation of the ascending aorta due to increased aortic blood flow, such as in tetralogy of Fallot, may compress the carina, left bronchus, and trachea.

Congenital cardiac disease accompanied by compression of the airways may result in a variety of clinical presentations, such as upper respiratory infection, wheezing, atelectasis, pneumonia, or hyperinflation. Incomplete obstruction can manifest with hyperinflation, while total obstruction results in atelectasis. Diffuse wheezing may occur as a result of extrinsic compression, bronchial narrowing, interstitial oedema, and airway hyperactivity.[10,17]

Imura et al.[6] demonstrated increased production of airway mucin in children who underwent cardiac surgery with cardiac pulmonary bypass. This was associated with the development of post operative complications such as collapsed lungs and pneumonia, and abnormalities in the exchange of gases, with an increased alveolar-arterial oxygen gradient. Several studies have suggested that increased glycoprotein and mucin concentration in the airways is associated with increased frequency of obstruction, bacterial infection, and abnormalities in the exchange of gases.[19,20]

This phenomenon might explain, in part, the high number of infections of the lower airways, and atelectasis, seen in our patients. Furthermore, external compression of the airways, atelectasis, and hyperinflation, even without increased production of glycoprotein and mucin in the airways, might interfere with excretions and movement of mucin in the airways, and its antibacterial and anti-infectious properties. Bacteria were isolated in only 3 out of 7 patients with evidence of an infectious process. This may be explained by the fact that most of the children were treated by broad spectrum antibiotics because of prolonged intubation. Prolonged extrinsic pulsation and compression of the airways may result in tracheobronchomalacia. The cartilaginous, muscular and elastic supporting structures of the airways are softer, weaker, and pliable in children, especially in infants, and therefore the airways are more prone to extrinsic compression and closure.[2,21,16] This collapse may result in complete occlusion of the lumen with functional abnormality and air trapping. Our fibreoptic bronchoscopies demonstrated tracheobronchomalacia in one-sixth of our patients, resulting in pathological air trapping, atelectasis and a very high peripheral airway resistance, which is inversely proportional to the radius of the lumen.

To the best of our knowledge, this is the first time that the rate of the procedures that resulted in alteration of treatment was discussed in the context of pulmonary involvement in children with congenital cardiac disease. Only occasionally is the bronchoscopic procedure itself curative. The relatively low rate of alterations in treatment might be explained by the high rate of spontaneous resolution of most of the pathologies found by us in the prolonged post operative period, such as tracheobronchomalacia.

When performing an invasive procedure, the potential complications should be taken into consideration, in particular in these very sick children. As with others, we encountered a very low rate of complications, only 6 in our 49 procedures, with 5 of these resolving immediately with supplementation of oxygen and lavage of the airways.[1,8] Only one patient required mechanical ventilation because of severe hypoxaemia. None of the patients died during or immediately after the procedure.

In conclusion, we believe that fibreoptic bronchoscopy has an important role in the evaluation of abnormalities of the airways in patients with congenital cardiac disease in whom rapid and accurate diagnosis is crucial for survival. Flexible bronchoscopy is safe, and contributed to a specific diagnosis in nine-tenths of the procedures we performed. We suggest, therefore, that fibreoptic bronchoscopy should be considered as an initial diagnostic tool in children with congenital cardiac disease and pulmonary involvement.

References

1. Lee SL, Cheung YF, Leung MP, Ng YK, Tsoi NS. Airway obstruction in children with congenital cardiac disease: assessment by flexible bronchoscopy. Pediatr Pulmonol 2002; 34: 304–311.

2. Corno A, Giamberti A, Giannico S, et al. Airway obstructions associated with congenital cardiac disease in infancy. J Thorac Cardiovasc Surg 1990; 99: 1091–1098.
3. deLorimier AA, Harrison MR, Hardy K, Howell LJ, Adzick NS. Tracheobronchial obstructions in infants and children. Experience with 45 cases. Ann Surg 1990; 212: 277–289.
4. Davis DA, Tucker JA, Russo P. Management of airway obstruction in patients with congenital heart defects. Ann Otol Rhinol Laryngol 1993; 102(3 Pt 1): 163–166.
5. Fischer JE, Allen P, Fanconi S. Delay of extubation in neonates and children after cardiac surgery: impact of ventilator-associated pneumonia. Intensive Care Med 2000; 26: 942–949.
6. Imura H, Duncan HP, Corfield AP, et al. Increased airway mucins after cardiopulmonary bypass associated with postoperative respiratory complications in children. J Thorac Cardiovasc Surg 2004; 127: 963–969.
7. Krastins I, Corey ML, McLeod A, Edmonds J, Levison H, Moes F. An evaluation of incentive spirometry in the management of pulmonary complications after cardiac surgery in a pediatric population. Crit Care Med 1982; 10: 525–528.
8. Chapotte C, Monrigal JP, Pezard P, et al. Airway compression in children due to congenital cardiac disease: value of flexible fibreoptic bronchoscopic assessment. J Cardiothorac Vasc Anesth 1998; 12: 145–152.
9. Lister G, Pitt BR. Cardiopulmonary interactions in the infant with congenital cardiac disease. Clin Chest Med 1983; 4: 219–232.
10. Berlinger NT, Long C, Foker J, Lucas Jr RV. Tracheobronchial compression in acyanotic congenital cardiac disease. Ann Otol Rhinol Laryngol 1983; 92: 387–390.
11. Bertrand P, Navarro H, Caussade S, Holmgren N, Sanchez I. Airway anomalies in children with Down syndrome: endoscopic findings. Pediatr Pulmonol 2003; 36: 137–141.
12. Green CG, Eisenberg J, Leong A, Nathanson I, Schnapf BM, Wood RE. Flexible endoscopy of the pediatric airway. Am Rev Respir Dis 1992; 145: 233–235.
13. Perez CR, Wood RE. Update on pediatric flexible bronchoscopy. Pediatr Clin North Am 1994; 41: 385–400.
14. Riedler J, Grigg J, Stone C, Tauro G, Robertson CF. Bronchoalveolar lavage cellularity in healthy children. Am J Respir Crit Care Med 1995; 152: 163–168.
15. de Blic J, McKelvie P, Le Bourgeois M, Blanche S, Benoist MR, Scheinmann P. Value of bronchoalveolar lavage in the management of severe acute pneumonia and interstitial pneumonitis in the immunocompromised child. Thorax 1987; 42: 759–765.
16. Fischer DR, Neches WH, Beerman LB, et al. Tetralogy of Fallot with absent pulmonic valve: analysis of 17 patients. Am J Cardiol 1984; 53: 1433–1437.
17. Kussman BD, Geva T, McGowan FX. Cardiovascular causes of airway compression. Paediatr Anaesth 2004; 14: 60–74.
18. Worsey J, Pham SM, Newman B, Park SC, del Nido PJ. Left main bronchus compression after arterial switch for transposition. Ann Thorac Surg 1994; 57: 1320–1322.
19. Aikawa T, Shimura S, Sasaki H, Ebina M, Takishima T. Marked goblet cell hyperplasia with mucus accumulation in the airways of patients who died of severe acute asthma attack. Chest 1992; 101: 916–921.
20. Bernstein JM, Reddy M. Bacteria-mucin interaction in the upper aerodigestive tract shows striking heterogeneity: implications in otitis media, rhinosinusitis, and pneumonia. Otolaryngol Head Neck Surg 2000; 122: 514–520.
21. Corno A, Picardo S, Ballerini L, Gugliantini P, Marcelletti C. Bronchial compression by dilated pulmonary artery. Surgical treatment. J Thorac Cardiovasc Surg 1985; 90: 706–710.