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Airway Complications after Single-Stage Unifocalization for Pulmonary Atresia, Ventricular Septal Defect, and Major Aortopulmonary Collateral Arteries

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Abstract

Objective: We analyze the incidence of postoperative severe airflow limitation after single-stage unifocalization in patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries (PA/VSD/MAPCAs) and comment on the treatment performed.

Methods: From 1994 until 2012, 106 patients with diagnosis of PA, VSD, MAPCAs underwent surgical treatment. Four of them (3.8%) developed in the postoperative course severe airflow complication. Chromosome 22q11 deletion was present in three of them. Median age at the time of unifocalization was 6.2 months (range 21 days – 11 months).

Results: The first developed malacia and compression of the left bronchus from the distal RV-PA conduit treated with external bronchial stenting with two incomplete costal cartilage rings. The second patient developed recurrent esophagus-left bronchus fistula treated with multiple surgical esophageal and bronchus reconstruction. The third child presented bilateral bronchial malacia treated with bilateral stenting and after with surgical elongation of the neo-left pulmonary artery to avoid external compression. The last developed bilateral bronchomalacia treated with bilateral bronchial stenting and after with RV-PA conduit replacement and endobronchial stenting calibration

Conclusion: Particular categories of patients (22q11 chromosome deletion, neonates/infants, patients with dominant/exclusive collaterals,) may be more predisposed to develop airway limitation. The treatment of the lesion should be individualized according to the supposed pathogenic mechanism. We suggest endoluminal treatment in absence of compression by vascular structures while surgery was used in proven case of extrinsic compression.

Keywords: Bronchomalacia; Major aortopulmonary collateral arteries; Pulmonary atresia with ventricular septal defect; Tetralogy of Fallot with pulmonary atresia

Introduction

The clinical outcome of patients with pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries (PA,VSD,MAPCAs) undergoing single-stage unifocalization can be complicated by airway problems occurring at different segments of tracheobronchial tree [1,2]. Besides the airway hyperresponsivness (AHR) typical of the disease and despite the standardization of surgical approach leading to achieve satisfactory and durable result [3,4], severe postoperative airflow limitation primarily related to airway malacia can contribute to significant morbidity and mortality [5]. After surgery patient can develop dependency from ventilation associated with respiratory distress unresponsive to medical management, requiring airway investigation and intervention. Etiology and pathogenesis of airway malacia associated to this disease and its surgical treatment is unknown, but different hypotheses have been suggested to give a plausible explanation [6]. Herein we report 4 cases of single-stage unifocalization in which postoperative malacia of the main bronchi occurred, in one patient in association with broncho-esophageal fistula.

Methods

From January 1994 until January 2012, 106 patients with diagnosis of PA, VSD, MAPCAs underwent single-stage unifocalization at our Institution. We reviewed the clinical and postoperative course of 4 of them (3.8%) who developed either in the early or late postoperative course severe airflow complication. Approval from the Institutional Scientific Board of the Bambino Gesù Children's Hospital was obtained. Median age at the time of unifocalization was 6.2 months (range 21 days – 11 months). All of them underwent a combination of angiocardiography, airway endoscopy, bronchography, and CT scan investigation showing malacia of the main bronchi, in one case in association with broncho-esophageal fistula.

The primary diagnosis and the demographic characteristics of our patient are reported in Table 1, instead the clinical course are illustred in Table 2.

N	SEX	Age	Genetic Diagnosis	Diagnosis	Aortich arch

1	F	11 mts	chromosome 22q11 deletion	PA, VSD, MAPCAs (3) severely hypoplastic and defective pulmonary arteries	left
2	F	6 mts	homozygous factor V Leiden	PA, VSD, MAPCAs (4), hypolastic and defective pulmonary arteries	right
3	M	21 days	chromosome 22q11 deletion	PA, VSD, absent pulmonary arteries, MAPCAs (4)	left
4	F	7 mts	chromosome 22q11 deletion	PA, VSD, severely hypoplastic pulmonary arteries, MAPCAs (6)	right

Table 1: Demographic characteristic and primary diagnosis of all children

mts: months; PA: pulmonary arteries; VSD: ventricular septal defect; MAPCAs: major aortopulmonary collateral arteries

N	Complication1	Procedure	Complication2	Procedure	Complication3	Procedure	Complication4	Procedure
1	Pseudo-aneurysm of distal anatomosis of the conduit	RV aneurismectomy with reconstruction of the left PA and replacement of RV-PA conduit	Compression of the left bronchus from the distal RV-PA conduit.	Removal of the bronchial compression by a shorter RV-PA conduit and external bronchial stenting with incomplete rings of costal cartilage				
2	Esophagus and left bronchus fistula	Esophageal direct suture and off-pump partial left bronchial resection with re-anastomosis	Recurrent esophagus-left bronchus fistula	Take down and reconstruction of confluence of PA, Directly sutures of esophagus, Re-anastomosed of the bronchus, Peribronchial omental wrapping	Respiratory distress syndrome	VA-ECMO (9 days)		
3	Neo-left PA stenosis al level of contiguity with the left bronchus	Extended reconstruction of the left PA	Bilateral bronchial malacia	Bilateral stenting of the main bronchi (dilated up to a caliber of 6 mm)	Left endobronchial stent ovalization,	Elongation of neo-left PA with tubular prosthesis	Ovalization of left endobronchial stent	PA confluence reconstruction and RV-PA conduit replacement
4	Diffuse bilateral bronchomalacia	bilateral bronchial stenting	Obstruction of SVC and stenosis of the left PA	Extended left PA reconstruction, SVC reconstruction, and RV-PA conduit replacement				

Table 2: Clinical Course of all patients of our study

PA: pulmonary arteries, RV: right ventricle, SVC: superior vena cavae, VA-ECMO: veno-arterial membrane oxygentation

Results

Case 1

An 11 month old infant with PA, VSD, MAPCAs (three), severely hypoplastic and defective pulmonary arteries, and chromosome 22q11 deletion underwent single-stage unifocalization and conduit right ventricular outflow tract (RVOT) reconstruction leaving the VSD open, due to an unfavorable intraoperative pulmonary flow study. Five months postoperatively, the child was readmitted at hospital for severe arterial desaturation and respiratory insufficiency. On echocardiography and cardiac catheterization a pseudo-aneurysm at the level of distal anatomosis of the conduit was demonstrated. The child was re-operated and a right ventricular aneurismectomy with reconstruction of the left pulmonary artery, and replacement of the right ventricle-pulmonary artery (RV-PA) conduit were performed. In the post-operative course, a total collapse of the left lung on chest-X ray was detected, in association with evidence of compression of the left bronchus from the distal RV-PA conduit on combined angiography and bronchoscopy. The child was then re-operated to remove the bronchial compression using a shorter and undersized RV-PA conduit. Surgical examination of the left bronchus showed complete malacia of its proximal portion: for such reason an external bronchial stenting with two incomplete rings trimmed out from costal cartilage was performed under intraoperative bronchoscopic guidance. Two years later the child underwent control cardiac catheterization and bronchoscopy: branching of reconstructed pulmonary arterial tree was still insufficient to allow VSD closure, owing to a Qp:Qs ratio less than 1. However, patency of left bronchus was satisfactory with a 25% reduction of its antero-posterior diameter at its origin. At the last out-patient clinical assessment, 3 years after last surgical procedure, the child is asymptomatic, with occasional tendency to arterial desaturation.

Case 2

A 6 month old infant with PA, VSD, MAPCAs (four), hypolastic and defective pulmonary arteries, and right aortic arch underwent

single-stage unifocalization and repair guided by an intraoperative pulmonary flow study. Soon after a fast-track postoperative discharge, the child was readmitted at casualty for fever and dyspnea, and further diagnostic investigation showed left lung pneumonia caused by aspiration due to a fistula developed between esophagus and left bronchus (Figure 1). The child was fasted and treated with parenteral nutrition and antibiotics for 12 days, afterwards she underwent surgical repair by esophageal direct suture and off-pump partial left bronchial resection with re-anastomosis performed through left thoracotomy. Three months later the child experienced recurrent left lung pneumonia related to recurrent fistula. This time an emergency procedure was required, performed through midline sternotomy on cardiopulmonary bypass. During this operation the confluence of reconstructed pulmonary arteries was taken down, the left main bronchus was transected at the level of the fistula, the esophagus was directly sutured, the bronchus was re-anastomosed, and a peribronchial omental wrapping was carried out to avoid further recurrence. The operation was completed with reconstruction of the pulmonary arterial confluence, and its early post-operative course was complicated by respiratory distress syndrome which was treated successfully with a 9 day course veno-arterial extracorporeal membrane oxygenation (VA-ECMO). At a follow-up interval of 10 months from the last surgical procedure, the child is asymptomatic; echocardiogram investigation shows good biventricular function with normal pulmonary arterial pressure; the left bronchus has a normal appearance on bronchoscopy.

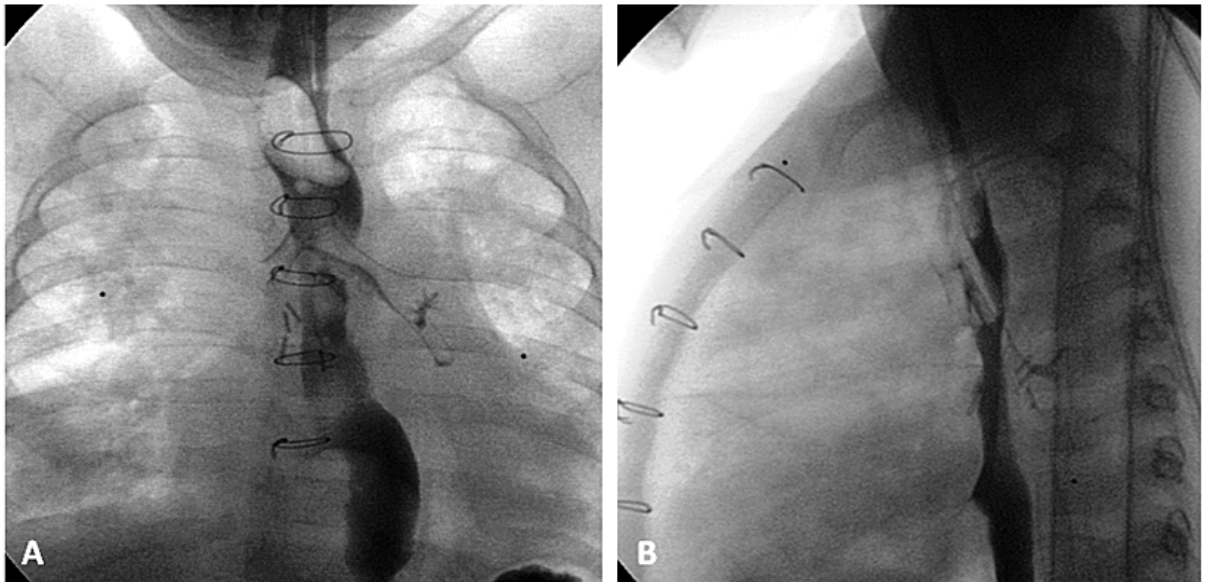

Figure 1: Esophageal contrastographic examination in anteroposterior (A) and lateral (B) projection showing passage of dye into the airway through a fistula between esophagus and left bronchus (Case 2).

Case 3

A 21 day old neonate with PA, VSD, absent pulmonary arteries, pulmonary blood supply provided by four MAPCAs, and chromosome 22q11 deletion underwent neonatal single-stage unifocalization and repair for severe respiratory and cardiac failure due to massive pulmonary overflow. Postoperative course was complicated by dependency from ventilator and a neo-left pulmonary artery stenosis in the area of anatomic contiguity with the left bronchus was diagnosed one month postoperatively owing to hypertensive right ventricular pressure. For this reason a new surgical procedure was performed consisting of an extended reconstruction of the left pulmonary artery which was folding below and behind the left main bronchus. For the persistence of respiratory insufficiency, both bronchoscopy and bronchography were performed, showing bilateral bronchial malacia (Figure 2A). It was treated with bilateral stenting of the main bronchi (Palmaz, Johnson and Johnson Interventional Systems, USA), which were dilated up to a caliber of 6 mm (Figure 2B). The child was eventually extubated and discharged home, but he came back a few months later for a worsening of respiratory function. A bronchoscopy showed left endobronchial stent ovalization, secondary to extrinsic compression by the neo-left pulmonary artery. A new surgical procedure of elongation of the neo-left pulmonary artery with a 7 mm PTFE tubular prosthesis was performed, followed one month later by a homograft RV-PA conduit replacement for fungal endocarditis of valve conduit. Five years later a new pulmonary arterial confluence reconstruction associated to conduit replacement was carried out for recurrent ovalization of the left endobronchial stent, together with a calibration of both endobronchial stents up to 10 mm. At the last out-patient clinical assessment, 1 year after the last surgical procedure, the child was asymptomatic, with normal airway patency.

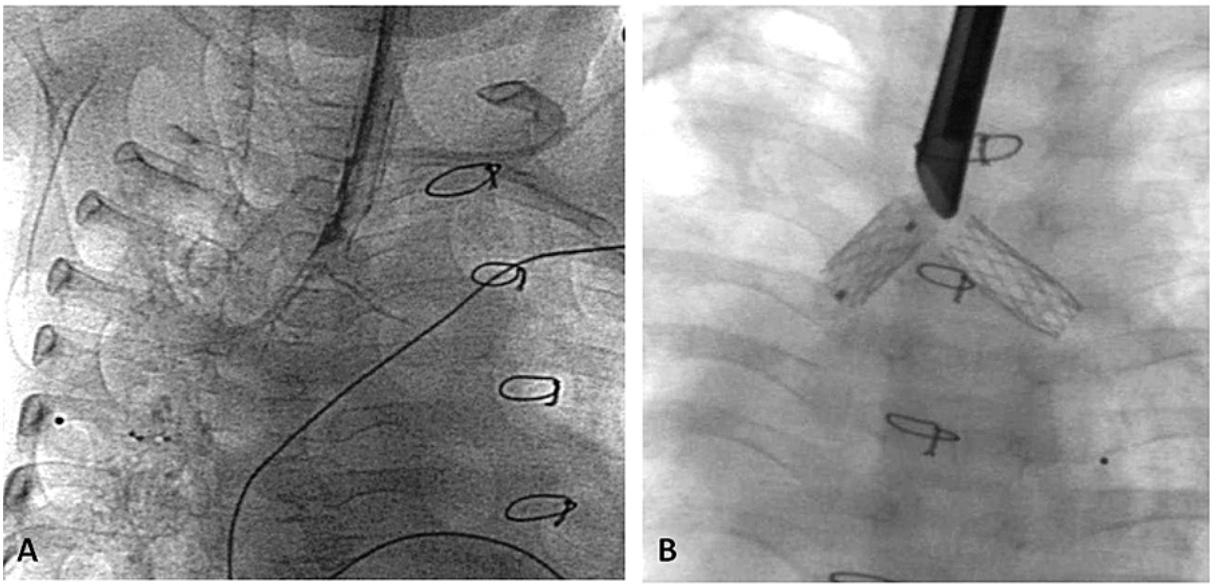

Figure 2: Bilateral malacia of main bronchi (Case 3). Contrastographic appearance (A) and result of bilateral stenting (B).

Case 4

A 7 month old infant with PA, VSD, hypoplastic pulmonary arteries, six MAPCAs, right aortic arch, and chromosome 22q11 deletion underwent single-stage unifocalization and repair guided by an intraoperative pulmonary flow study. In the post-operative course she received tracheostomy due to prolonged intubation, and subsequently bilateral bronchial stenting due to the evidence of diffuse bilateral bronchomalacia on bronchoscopy. Two years later, a new diagnostic assessment showed obstruction of superior vena cava and stenosis of the left pulmonary artery, treated with extended left pulmonary artery reconstruction, superior vena cava reconstruction, and RV-PA conduit replacement. Endobronchial stenting calibration was routinely yearly performed up to a 10 mm size. At the follow-up interval of 5 years from the last surgical procedure (approximately seven years from endobronchial stenting) the patient showed a reduced functional capacity associated with dilated hypertensive right ventricle. Nevertheless, airway patency was absolutely satisfactory.

Discussion

The development of bronchial or tracheal malacia in patients undergoing treatment for PA, VSD, MAPCAS is generally associated with poor prognosis [5]. In spite of the scarce knowledge about etiology and pathogenesis of such lesion, several hypoteses can be made to justify its occurrence.

Chromosome 22Q11 deletion

PA, VSD, MAPCAS is typically associated with chromosome 22q11 deletion [4,7-9], and, as reported in several papers [10-12] this association can be sometimes affected by airway abnormalities as a result of external vascular compression. Therefore the occurrence of chromosome 22Q11 deletion in 3 patient (75%) of our series is not surprising.

External compression

The origin of MAPCAs from the descending aorta may cause constriction of the space between ascending and descending aorta and the aorto-spinal distance, creating the anatomic condition for a compression of the tracheobronchial tree posteriorly by the descending aorta and anteriorly by the MAPCAs [10,13]. Two patients in our group had also right-position of the aortic arch: this condition may further contribute to airway compression. Another possible explanation was suggest by McElhinney et al. [1] who reported an increased susceptibility to compression in the infant tracheobronchial tree due to the small bronchial caliber associated with lose of resilience of young airway that remain prone to malacia despite surgical treatment. As a matter of fact, all patients of our series were either neonates (one) or infants (three).

Intrinsic pathological changes of infant airway

Besides external anatomic compression, intrinsic pathological changes of infant airway might be advocated as a potential cause of malacia in relationship with anatomic abnormalities of bronchial vascularization and/or bronchial blood supply. In fact, the airway hyperresponsivness (AHR) typical of such disease is responsible for an alterated bronchial tone that may cause abnormal bronchopulmonary circulation and hemodynamics.

The result would be a weakness of the bronchial wall, which would be further enhanced by the ischemia-reperfusion sequence related to the surgical procedure of unifocalization. Schulze-Neick et al. [5] reported three patients who developed airflow limitation after unifocalization and eventually died. On postmortem severe tracheobronchial extensive necrosis of ischemic origin was demonstrated in all cases, suggesting that the nutritive supply to airway epithelium was probably already compromised before surgery. Finally, abrupt changes of the bronchial perfusion associated to the manipulation of collaterals during surgery may be advocated as a potential cause of postoperative malacia. In according with other

authors [14,15], we consider the MAPCAs as dilated bronchial arteries. Indeed it is undeniable, for example, that the nutrient arteries of the lymphnodes arise from MAPCA and during one-stage unifocalization performed in patient with exclusive MAPCA, the bronchial circulation is almost completely abolished as demonstrated by the complete absence of blood back into the collaterals in the course of surgical manipulation.

Then we can assume that the MAPCAs are highly responsible for the airway perfusion and may postulate that any interruption or significant reduction of their blood supply may result in weakening of bronchial wall. At the same time, the successful use of MAPCAs as functional pulmonary circulation does not allow to exclude the potential possibility to produce bronchial ischemia, which may be much more extensive in patients with exclusive MAPCAs due to the absence of the so-called retrograde bronchial collateral circulation of pulmonary origin.

It has already been reported that collateral obliteration may cause airway wall ischemia and weakening due to abrupt interruption of bronchial arterial supply [1]. Surgical unifocalization of collaterals, while not abolishing the bronchial arterial perfusion, would lead to a drastic reduction of their perfusion pressure, by shifting their function of nutrient arteries in that of functional vessels. This effect would be especially evident in the case of dominant MAPCAs, even more if hypertensive prior to unifocalization, particularly in the case of simultaneous VSD closure. All patients of our study had either dominant or exclusive MAPCAS; three of them underwent single-stage unifocalization and simultaneous VSD closure, and the airway complication was probably due to ischemia as a consequence of hypoperfusion. The ischemia can be discrete o diffuse according to extent of the collateral circulation unifocalized.

It will possibly be more relevant (bi-bronchial for example) in patients with exclusive MAPCAs, as in the case 3. It may probable be restricted in patients with pulmonary circulation not exclusively provided by MAPCAs, due to the effect of so-called retrograde bronchial collateral circulation of pulmonary origin (namely the same circulation which enables the consolidation of the bronchial anastomosis in lung transplantation). In one case only the VSD was left open, due to unfavorable intraoperative pulmonary flow study, and the airway malacia was probably related to compression of tracheobronchial tree by the RV-PA conduit. Putting aside the case of broncho-esophageal fistula that required surgical resection and bronchial reanastomosis [16], performed twice, the approach to airway malacia was different depending on whether bronchial compression had been detected or not. In the absence of compression by vascular structures, endoluminal treatment was privileged, while surgery was used in the proven case of extrinsic compression. In the latter case, in addition to removing the compression produced by the conduit, we decided to externally stent the left bronchus using incomplete rings of costal cartilage [17] in order to avoid the potential for erosion of an endobronchial stent on the contiguous vascular structure.

In all cases interventional treatment of bronchomalacia was successful, although endoluminal stenting required repeated calibration procedures consensual to patient growth. Finally, endoluminal stent patency was reduced twice in one case due to left main bronchus compression by an anomalous course of the left pulmonary artery. Left pulmonary artery elongation prior to stent dilation was performed, in order to avoid any potential for stent erosion.

Conclusion

Bronchomalacia requiring investigation and treatment may complicate the postoperative course after unifocalization for PA, VSD, MASPCAS. Overall incidence is low, although particular categories of patients (carriers of 22q11 chromosome deletion, neonates/infants, patients with pulmonary vascularity pattern characterized by dominant/exclusive collaterals, especially if hypertensive) may be more predisposed to develop this type of complication. Despite the fact that etiology and pathogenesis remain unclear, different mechanisms may be postulated related to the disease itself and to the unifocalization procedure. There is really no way to prevent or predict the likelihood of its postoperative development. In case of occurrence, the treatment of the lesion should be individualized according to the supposed pathogenic mechanism, and it may lead to satisfactory medium term results.

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