Images in cardiovascular medicine
Life-threatening hemoptysis after the Fontan procedure

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A 21-year-old patient with complex congenital heart disease was referred to our unit due to recurrent life-threatening hemoptysis. He had pulmonary atresia with an intact ventricular septum and had been submitted to modified left and right Blalock-Taussig shunts at birth and at the age of 7 years respectively. He had subsequently been submitted to a bidirectional cavo-pulmonary anastomosis and to a modified Fontan procedure with the positioning of an intracardiac conduit at the ages of 13 and 16 years respectively. Mid-term follow-up controls after the Fontan operation did not reveal any significant anomaly. Ten days before hospitalization, due to a moderate self-limited hemoptysis, the patient had been submitted elsewhere to chest X-ray and computed tomographic scan that revealed partial atelectasis of the lower lobe of the right lung. Then, a new episode of severe bleeding from the upper airways prompted the immediate referral to our department. At admission, the patient was in fairly good clinical conditions but for a mild dyspnea. At bronchoscopy, a huge organized clot, totally occluding the right main bronchus, was found (Fig. 1). It was mechanically lysed and removed during bronchoscopy, so relieving the airway obstruction. At the follow-up bronchoscopic control performed a few days later, some dilated and tortuous anomalous arterial vessels protruding into the right bronchial lumen were imaged (Fig. 2A). They branched off into multiple tufts of capillaries that formed a fragile capillary network within the bronchial mucous membrane (Fig. 2B). In order to search for, and possibly occlude, these anomalous vessels, cardiac catheterization was performed.

At cardiac catheterization, no vascular anomaly of the pulmonary tree (i.e. arterial malformations or artero-venous fistulas) was imaged (Fig. 3). Conversely, an anomalous dilated, tortuous bronchial artery arising from the upper thoracic aorta was found (Fig. 4A). It wrapped around the proximal segment of the right bronchus giving rise to an extensive capillary network that presumably was the source of the episodes of massive bleeding. Then, the feeding source of this anomalous vascular network was selectively injected and occluded with multiple stainless steel spring coils (Fig. 4B). At 6 months of follow-up the patient no longer complained of any further episodes of airway bleeding.

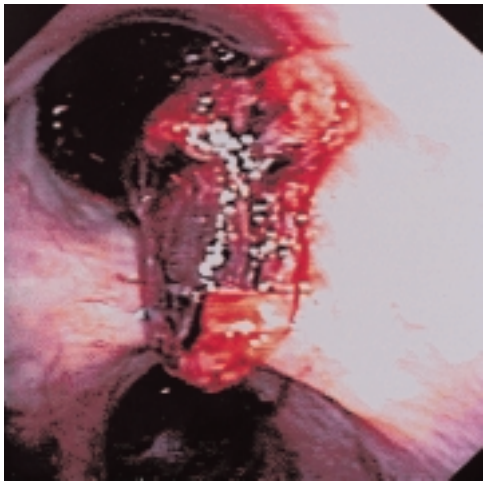

Figure 1. *Bronchoscopic image of a huge organized clot almost completely occluding the right main bronchus and resulting in partial atelectasis of the right lung.*

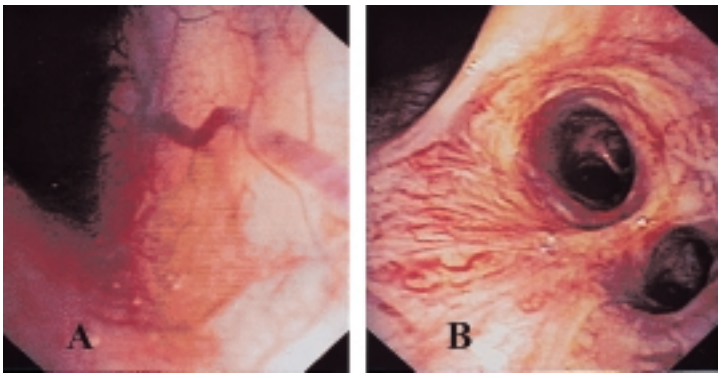

Figure 2. *Control bronchoscopy, performed a few days after lysis and removal of the clot, that shows a long, tortuous artery that entwines the proximal part of the right bronchus* (A) *and gives rise to a fragile submucosal capillary network* (B).

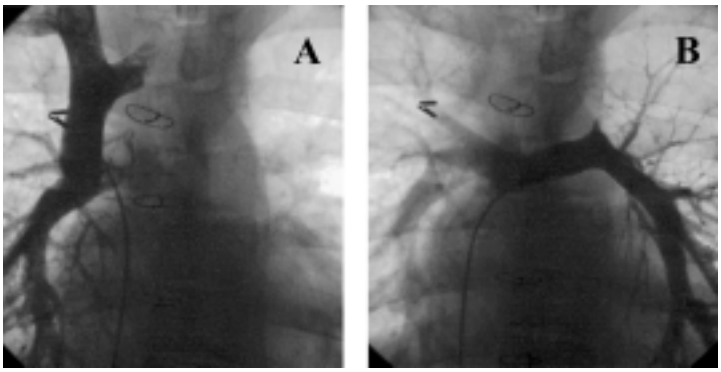

Figure 3. *Right* (A) *and left* (B) *pulmonary angiography performed via the cavo-pulmonary anastomosis and showing a normal peripheral distribution of the arterial tree without any vascular anomaly potentially causing the airway bleeding.*

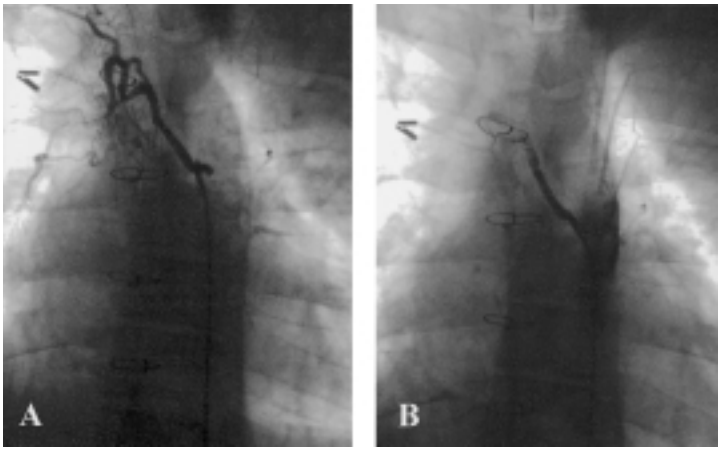

Figure 4. *Selective injection into an anomalous bronchial arterial vessel originating from the upper thoracic aorta and giving rise to a capillary network around the proximal right bronchus, before* (A) *and after* (B) *the endoluminal embolization with multiple stainless steel spring coils.*

Bronchial artery enlargement is a well-known long-term complication of congenital heart disease with a reduced total[1,2] or effective[3] pulmonary blood flow and, in case of an unbalanced pulmonary perfusion, it is frequently found in the less perfused lung. It could be speculated that this was true even in our patient who had an unbalanced pulmonary blood flow through the left Blalock-Taussig shunt for a long time. Bronchial artery enlargement frequently causes life-threatening hemoptysis, the treatment of which is still very challenging[2,4,5]. Percutaneous embolization is now considered a safe and effective treatment even in critical patients, with a better cost-effective ratio compared to other medical or surgical therapeutic options. In our patient, the transcatheter embolization with multiple stainless steel spring coils of the sole bronchial artery

malformation imaged at aortic angiography dramatically improved the clinical conditions and, hopefully, definitively eliminated the risk of further episodes of bleeding.

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