Case Report

Large aortopulmonary collaterals: a cause of respiratory failure or severe bronchitis

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Abstract

Two infants presented with recurrent lung disease early in life. One was premature with bronchopulmonary dysplasia and oxygen dependent. The other had two episodes of pneumonia/severe bronchitis necessitating hospital admission at one and three months. In both of them multiple aortopulmonary collaterals were diagnosed and coil occlusion of these collaterals led to improvement of the clinical status in both of them.

Introduction

Lung disease in early infancy can be attributed to different causes. Aortopulmonary collaterals have rarely been described as a cause for chest problems in premature infants.[1] We present two infants in whom aortopulmonary collaterals were identified as one of the major causes of their respiratory symptoms and occlusion of these in collaterals led to improvement of the clinical condition both of them.

Case Report

Case 1: First case was a male child, born in another institution, at 26th gestational week. He was treated with Indomethacine for patent ductus arteriosus. He developed severe bronchopulmonary dysplasia and remained oxygen dependent for three months when for the first time he was seen at King Faisal Specialist Hospital and Research Centre.

On examination he appeared in good health, his weight was 2.74 kg and height was 46 cms, oxygen saturation of 95% on 50% of Oxygen by mask, palpable peripheral pulses, no organomegaly, hyperactive precardium and a systolic murmur over the left sternal border. The echocardiography showed multiple aortopulmonary collateral arising from arch of aorta. No patent ductus arteriosus or other cardiac defect could be seen. The best visualization for the collaterals was from the suprasternal view. By color Doppler the collaterals will have the appearance of a patent arterial duct, but in contrast they will not open into the pulmonary artery as the patent arterial duct will do. Of course, the examiner should think of this possibility when she/he is doing the echocardiographic study.

We speculated that reducing left to right shunt to the lung would reduce lung congestion and edema and hence could improve clinical situation of the patient. Therefore, cardiac catheterisation was performed to close the collaterals.

Right heart catheterisation showed a slightly elevated pulmonary artery pressure. Aortography confirmed multiple aortopulmonary collateral arising from lower thoracic aorta and supplying lower lobe of the right lung. There was no patent ductus arteriosus or coarctation of the aorta. One, very large collateral (diameter of 4 millimetre), draining the right lower lobe was successfully occluded using a 5 mm x 5 cm Gianturco coil (Figure 1a, b). The intervention was uneventful. All pulses were palpable after the procedure.

Within 24 hours, the oxygen was weaned off and he was discharged on 5th day, on oral feeding.

He was followed up in the clinic at one month (weight=4.62 kilograms, height=54 centimetres) and six months (weight=8 kilograms, height=79 centimetres) after the procedure. The oxygen saturation was more than 93% on room air and echocardiogram still showed at least two but, tiny collaterals.

Case 2: The second patient was admitted twice in early infancy to this hospital with severe respiratory symptoms. The first admission at one month was because of a para-hilar right- sided pneumonia. Echocardiography, done because of a systolic murmur, demonstrated multiple aortopulmonary collaterals. A tiny muscular ventricular septal defect was also visualised. Chest infection was treated and was discharged.

The second admission at three months was again due to respiratory symptoms. Examination showed weight=5.52 kilograms, (50-75% centiles), height=57 centimetres (25-50% centiles), heart rate=130/minute and respiratory rate=62/minute. Severe bronchospasm necessitated oral brochodilator Bricanyl, Prednisone and Pulmicort nebulisation.

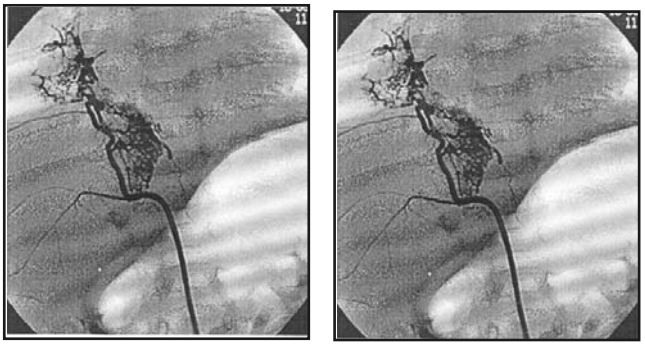
Echocardiography showed spontaneous closure of the muscular ventricular septal defect. One major aortopulmonary collateral, 3 mm in diameter, was still visualized. Cardiac catheterisation was performed one month later at 4 months of age (weight=6 kilograms, height=64 centimetres). Haemodynamic data showed normal Pulmonary artery pressure. Aortography demonstrated multiple aortopulmonary collaterals draining to both (mainly right) lungs. The pressure in the largest collateral was systemic. The collateral to the right lung was successfully occluded using a 5 millimetres X 5 centimetres Gianturco coil (Figure 2a, b) as a day case procedure and discharged a few hours after the intervention.

She was followed up one and seven months after the intervention and according to the parents, she had only one brief episode of bronchitis since occlusion of the collaterals, which did not necessitate hospital admission and no collaterals were seen on echocardiography.

Discussion

Lung disease in premature babies and during early infancy can be attributed to different causes. Aortopulmonary collaterals have occasionally been described as leading to congestive heart failure necessitating trans-catheter treatment.[1] In this report, aortopulmonary collaterals were demonstrated by Doppler echocardiography in 66% of the studied premature babies. While 11% of these patients were treated for congestive heart failure, only one underwent occlusion of a major collateral using embolization coils. Sign of left to right shunt were extrapolated indirectly from the dilated cardiac silhouette, the increased vascularity on the side of the collaterals in the chest radiograph, as well as clinically. According to our knowledge these collaterals were not described as a major cause for respiratory failure in early infancy (i.e. oxygen dependency or bronchitis/brochiolitis).

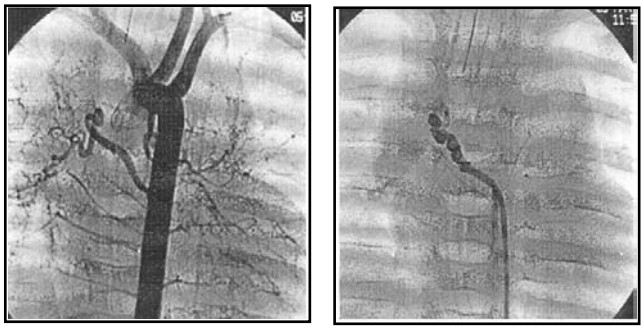
There are different vascular abnormalities, which can lead to recurrent chest infections[2] and can give similar Colour Doppler imaging on echocardiography.[3] According to our experience the best visualization for the collaterals was from the suprasternal view. By color Doppler, the collateral will have the appearance of a patent arterial duct, but in contrast they will not open into the pulmonary artery as the patent arterial duct will do. Besides more common lesions as patent ductus arteriosus, there are relatively rare anomalies like aortopulmonary window, aortopulmonary fistulas and bronchopulmonary sequestration. Aortopulmonary fistula is usually a fatal condition with haemoptysis, if not treated early.[4] The most frequent cause is erosion of a false aneurysm of the descending thoracic aorta into the left lung. The patients would have had a history of haemoptysis, a previous thoracic aortic surgical procedure, or a thoracic aortic aneurysm.



1a. **1b.**

Fig 1a,b. Selective angiography in the collateral supplying the lower lobe of the right lung before (1a) and after (1b) the procedure showing complete occlusion.

There are two types of bronchopulmonary sequestrations. The intra-lobar sequestration share the same visceral pleura as a normal lung and their venous drainage is normal. The extra-lobar sequestrations have distinct pleura from the rest of the lung and their pulmonary veins drain into the azygous system.[5] Lung sequestrations are usually fed by collaterals derived from the abdominal aorta. In our case the collaterals were multiple and were derived from the thoracic aorta indicating that these were not feeding on a sequestered part of the lung.



2a. **2b.**

Fig 2a,b. Angiography in the descending aorta before (2a), and selective angiography in the occluded collateral after (2b) the procedure, showing multiple aorto pulmonary collaterals with one major collateral directed towards the middle lobe of the right lung and complete occlusion after successful deployment of one 5 cm X 5mm Gianturco coil.

Embryologically, the central pulmonary artery is formed by the sixth branchial arch at the 32nd day of gestation. Before this event, paired segmental arteries arising from the dorsal aorta supply the lung buds. Within 50 days of gestation these arteries normally would have regressed.[6]

In our case we postulate that the collaterals occluded are segmental arteries, which did not undergo spontaneous regression. There is a possibility that these might also be collaterals which "proliferate in response to a given stimulus" i.e. as a result to long-term mechanical ventilation.[1] Hemodynamically, they lead to congestive heart failure due to excessive left to right shunt, and pulmonary vascular obstructive disease of the affected lung segments. This was the cause in one patient with pulmonary oedema and oxygen-dependency and in the other, the main reason of recurrent chest infections. Though the detailed precoil status was not quantified, we believe that the beneficiary impact of coil occlusion on these two patients were shown by the fact that the first child could be weaned off oxygen within 24 hours after the occlusion, and that the 2nd child was not hospitalized over the length of the study period.

On the basis of these cases, we suggest that premature babies on positive pressure ventilation, and the children with recurrent lung infections during early infancy, who have clinical suspicion or radiological evidence of aortopulmonary collateral or other left to right shunts, should routinely have detailed echocardiographic examination including Colour Doppler study to rule out aortopulmonary collaterals as a possible additional cause for the lung disease. Occlusion of these collaterals can improve the clinical status of the patients.

References

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