Case Report

"Asthma": An Unusual Presentation of Cor Triatriatum

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We discuss an extremely unusual presentation of a 19-month-old child with *cor triatriatum* and an intact interatrial septum, who presented for the first time at the age of 16 months with wheezing and repeated lower respiratory tract infections. At surgery, a thick fibromuscular membrane with a 2-3 mm eccentrically placed orifice was identified, and following surgical resection of the membrane the child made an uneventful recovery. This case demonstrates the need for investigating children with "asthma" who do not respond to conventional medical management. A rare but potentially correctable underlying cause may be found.

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Cor triatriatum is a rare congenital cardiac anomaly, accounting for 0.1% of all congenital cardiac malformations, and is characterised by the presence of a membrane dividing the left atrium into two chambers. These children usually present at a very young age with cardiac failure and repeated lower respiratory tract infections (LRTI).

Case presentation

A 19-month-old male presented to the paediatricians at a district general hospital with a three-month history of recurrent LRTI and wheezing. Following routine investigations, the child was diagnosed as having asthma and commenced on bronchodilator therapy. In view of the severity and frequency of these episodes he was given nebulisers and oral steroids. However, his symptoms recurred and resulted in four admissions to hospital over a period of three months. Renal and liver function tests were normal, while blood gas analysis revealed a metabolic acidosis.

On his last hospital admission, plain chest radiography was performed and revealed mild cardiomegaly with signs of pulmonary congestion. The electrocardiogram was suggestive of right ventricular hypertrophy (RVH). Hence, the child was referred to cardiologists at our centre with an initial diagnosis of pulmonary hypertension.

An echocardiogram performed on admission revealed right atrial enlargement with RVH and tricuspid regurgitation, suggestive of pulmonary hypertension. In addition, a thick membranous structure was noted in the left atrium, separating it into two chambers. This had an eccentrically placed orifice nearer to the septal margin of the membrane (Figures 1A, B). In the absence of any other structural abnormalities, a diagnosis of *cor triatriatum* was made and the findings of pulmonary hypertension were attributed to it. A decision was made to proceed to urgent corrective surgery.

In the operating room, prior to institution of cardiopulmonary bypass (CPB), the child showed evidence of worsening pulmonary hypertension, in the form of severe haemodynamic instability with rising pulmonary arterial pressures associated with a severe degree of desaturation. This requir-

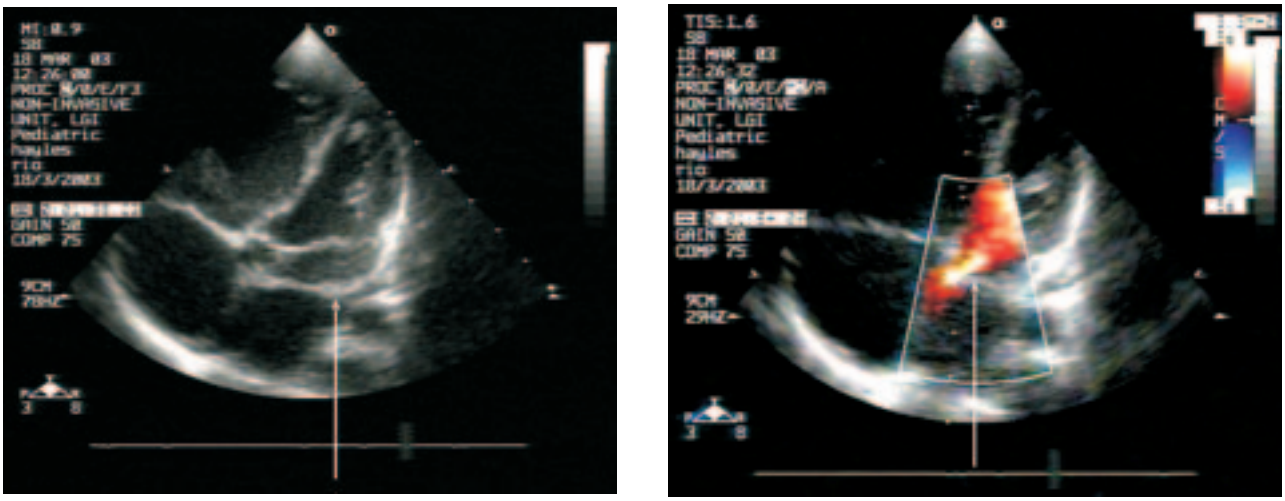

Figure 1. A: Preoperative echocardiography demonstrating the *cor triatriatum*. The arrow indicates the membrane dividing the left atrium into two chambers. B: Preoperative Doppler echocardiography demonstrating the *cor triatriatum*. The arrow indicates blood flow through the narrow orifice in the atrium.

ed ventilatory manipulation to achieve haemodynamic stability. CPB was rapidly instituted with aortic and bicaval cannulation. The acidosis improved after institution of CPB. The patient was cooled down to 28° C. The heart was arrested with cold, antegrade, crystalloid cardioplegia and the right atrium was opened. An incision was then made into the intact interatrial septum and the membrane was visualised. All four pulmonary veins were seen to drain into the proximal chamber and the two chambers were separated by a thick fibromuscular membrane with a 2-3 mm eccentrically placed orifice (Figure 2A). The membrane was completely excised and the anatomy of the left atrium was inspected (Figure 2B). The mitral valve was normal. The raw area on the wall of the left atrium was oversewn with 5-0 prolene sutures. The incision in the atrial septum was patched with autologous pericardium and the child was weaned off CPB without problems, following the closure of the right atriotomy. The child made an uneventful recovery following surgery and was discharged home on the sixth post-operative day.

Discussion

Cor triatriatum is an extremely rare congenital anomaly, characterised in its classical form by a fibromuscular

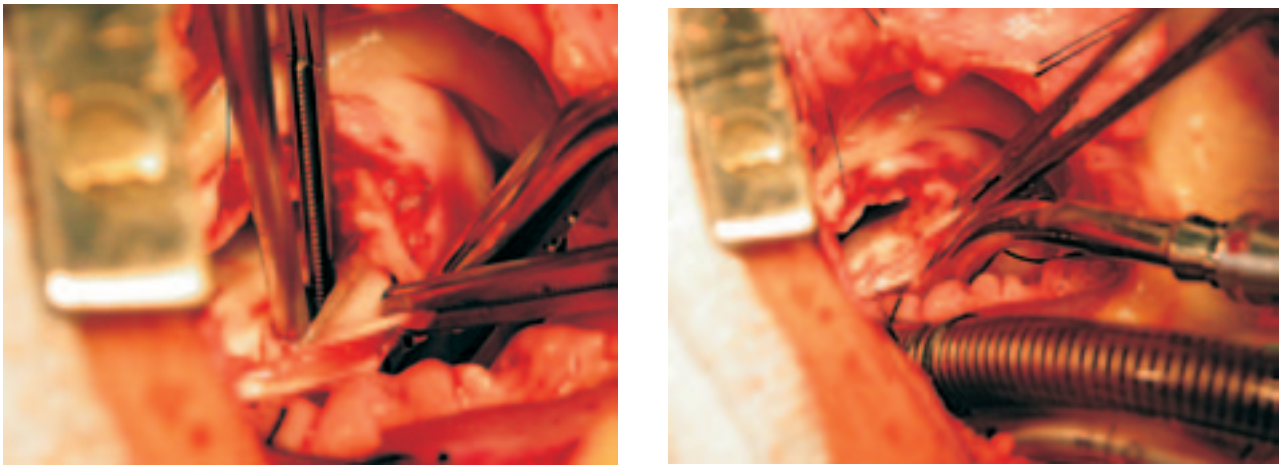

Figure 2. A: Intraoperative photograph showing a dissecting forceps through the orifice in the membrane. B: Intra-operative photograph after the excision of the membrane.

membrane with an orifice of varying diameter, separating the left atrium into two chambers. The pulmonary veins often drain into the proximal chamber. *Cor triatriatum* is often associated with other cardiac congenital anomalies, such as atrial septal defect and partial anomalous pulmonary venous drainage. More rarely, it is seen in association with tetralogy of Fallot and complete transposition of the great arteries.

The clinical presentation depends on the degree of obstruction to the blood flow from the upper to the lower chamber and the presence of associated congenital anomalies.[1] Most patients present in the neonatal period or early infancy with a failure to thrive and cardiac failure. These patients usually have a narrow orifice in the membrane between the two chambers and an interatrial connection. There have been a few cases reported in adulthood, but these are usually picked up as incidental findings on cardiac imaging or present with gradually worsening shortness of breath.[2,3] The orifice connecting the two chambers is usually much larger in adults and children presenting later in life. An association with an atrial septal defect or a patent *foramen ovale* is seen in up to 80% of cases.[4]

The case described here is unique, as it is an unusual and delayed presentation of *cor triatriatum* with a very narrow orifice in the membrane between the two chambers. The only presenting symptom in this child was recurrent LRTI and wheezy episodes starting at the age of 15 months. Interestingly, one such case of *cor triatriatum* in a child presenting with wheezing the sole symptom has been reported previously.[5] It is not clear whether that child had an intact atrial septum. However our patient had remained completely asymptomatic for the first 15 months of his life, despite having a very narrow orifice in the membrane between the two chambers and an intact atrial septum. Of particular note is the observation that the child did not respond to bronchodilators and steroids as would have been expected if he actually had bronchial asthma.

His rapid and continuous deterioration prompted urgent investigations and surgery. His recovery following surgery was remarkably uneventful. This case also reveals the importance of an early and accurate diagnosis of *cor triatriatum* in such unusual circumstances, as it is amenable to curative surgery. This would in turn prevent the long-term sequel of pulmonary hypertension and result in a better prognosis.

Pulmonary hypertension as a result of elevated left-sided pressures usually indicates the severity of the lesion, and worsening pulmonary hypertension is associated with a poor prognosis. These children are extremely susceptible to severe pulmonary hypertensive crisis, especially in the face of any form of stress. In this particular case, the child suffered a crisis in the operating theatre just after induction.

In conclusion, this case illustrates an unusual presentation of *cor triatriatum* and the importance of investigating for less common causes of wheeze and chest infections in children, especially those who do not respond to appropriate treatment. Timely surgical intervention for potentially curable lesions is associated with better outcomes.

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