Cardiol Young 2002; 12: 491–493

ISSN 1047-9511

Brief Report

Rapidly progressive idiopathic dilation of the right atrium in infancy associated with dynamic obstruction of the airways

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Abstract We present a young infant with rapidly progressive idiopathic dilation of the right atrium, complicated by symptomatic and dynamic obstruction of the airways secondary to proximal obstruction which was reversed subsequent to surgical reduction of the right atrium. We discuss the surgical implications of the distorted cardiac anatomy, along with post-operative complications, which included refractory arrhythmias.

Keywords: Atrial dilation; bronchial obstruction; congenital heart disease

Idiopathic dilation of the right atrium is a rare congenital anomaly, first described by Bailey in 1955.[1] Many patients are asymptomatic, being discovered incidentally, while others present with arrhythmias, fatigue, or shortness of breath, usually at an older age.[2] Optimal management of those patients who are asymptomatic remains controversial.

Case report

S.H. was diagnosed to have massive right atrial dilation and moderate tricuspid regurgitation during fetal echocardiography at 35 weeks gestation. There was no evidence of Ebstein's malformation of the tricuspid valve. The degree of right atrial dilation was thought to be out of proportion to the degree of tricuspid regurgitation. The baby delivered via normal spontaneous vaginal delivery. Initially, the baby had mild tachypnea and minimal cyanosis, attributed to echocardiographically documented right-to-left shunting across the atrial septal defect, which resolved in a few days. The chest radiograph was significant for cardiomegaly. Postnatal echocardiography illustrated massive right atrial dilation, with the chamber measuring 55 mm by 35 mm, which was out of proportion to the degree of tricuspid regurgitation. There was no evidence of Ebstein's malformation, but the tricuspid valve appeared dysplastic. The tricuspid annulus was measured at 10.5 mm. There was a small atrial septal defect. The right ventricle was normally formed, and of normal size, with neither inflow nor outflow obstruction. The left heart structures were also normal. The baby was able to feed well, maintain adequate systemic oxygen saturation, and was discharged from the neonatal intensive care unit.

The baby was re-admitted to the hospital for a possible viral infection with some increased work of breathing and episodic cyanosis at two weeks of age. Repeat cardiac evaluation was not significantly different, and the patient was in normal sinus rhythm. The right atrium now measured 57 mm by 45 mm. In the first 4 weeks after birth, there was rapid dilation of the right atrium to 70 by 50 mm. Over the next 4 weeks, there was progressive enlargement with significant displacement of the cardiac structures, such that the patient had to be rolled into right lateral position to obtain echocardiographic views. It was no longer possible to measure the right atrial dimensions, as the right atrium wrapped itself around the great vessels and the major systemic veins.

During hospitalization, the patient continued to have episodes of respiratory distress mimicking severe bronchospasm, and these episodes were associated with cyanosis. Computerized axial tomography of the chest did not identify any discrete obstruction within the

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Accepted for publication 10 June 2002

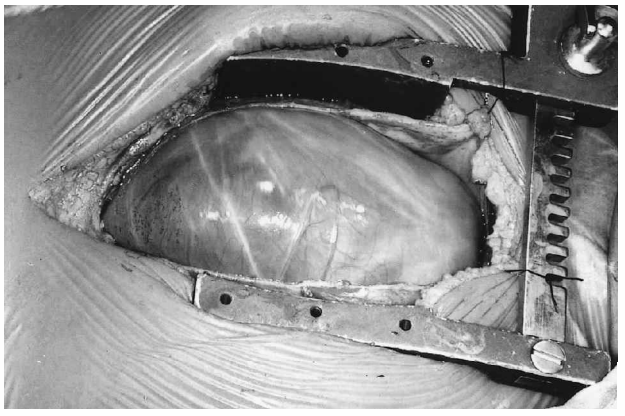

Figure 1.
The tense and dilated right atrium as initially seen bulging through the sternotomy on opening of the pericardium. The head of the patient is to the right of the picture.

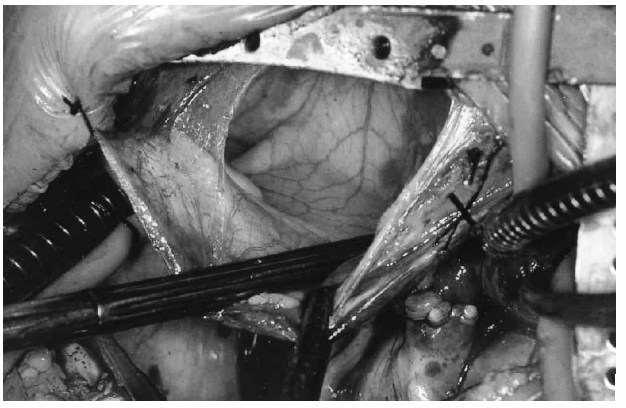

Figure 2.
Having opened the massively dilated right atrium, its extremely thin, nearly translucent, walls are evident. The head of the patient is again at the right of the picture.

airways, although fluoroscopy showed dynamic distal collapse during agitation and crying.

At this time, two options were considered in the absence of any guidelines from review of the literature. The first was to carry out further investigations and tracheostomy to treat possible tracheobronchial pathology, invoking a different diagnosis for the respiratory symptoms and monitoring progress of right atrial dilation. Alternatively, we could surgically reduce the volume of the right atrium, making the assumption that right atrial dilation was responsible for the respiratory manifestations. If the patient continued to have respiratory symptoms and failed extubation after surgery, then additional investigations and possibly tracheostomy would be performed to treat possible tracheobronchial disease. There was additional concern that the size of the right atrium was interfering with growth of the right lung.

After deliberation, we opted for the second course of action. Bronchoscopy performed at the time of the operation showed mild proximal tracheomalacia. The rest of the airway was unremarkable. After sternotomy, and pericardial opening, the right atrium was seen to be bulging out of the sternotomy (Fig. 1). The right atrium was noted to be massively dilated, occupying virtually the entire mediastinum, and was compressed behind the sternum. The superior and inferior caval veins were obscured from sight by the right atrium, and the aorta was displaced posteriorly approximately 6 cm from the sternum. The atrial wall was thin, fibrotic, and appeared non-functional (Fig. 2). The tricuspid valve appeared dysplastic, but there was no evidence of Ebstein's malformation. The right coronary artery was displaced 2 cm laterally from its normal position, stretched over the dilated atrium, and was obscured by the fibrosis on the atrial surface. Reduction atrioplasty was performed, resecting all but a rim of approximately 2 cm of the right atrium, which was then closed directly. In the process, the stretched right coronary artery was inadvertently excised. This was primarily repaired. The atrial septal defect was closed. The pericardium was tacked to the lateral wall of the atrium with a running suture, to act as a buttress supporting the remaining thin wall of the right atrium.

The infant was easily weaned from cardiopulmonary bypass with good biventricular function, and was successfully weaned from assisted ventilation postoperatively without difficulty. Postoperative fluoroscopy of the airway was normal. The patient had complete resolution of previously described episodic respiratory symptoms and cyanosis.

The initial postoperative period was complicated by episodes of junctional ectopic tachycardia and supraventricular tachycardia. A few weeks later, the patient developed fairly refractory atrial tachyarrhythmias requiring treatment with amiodarone. This has resulted in suppression of the arrhythmia, which now persist only as non-sustained runs.

Discussion

Several unusual features were encountered during the care of this patient, some of which have not been reported in literature. These include rapid progression of right atrial dilation within a span of 8 weeks, respiratory compromise presenting as episodic increased work of breathing with cyanosis, reversible with atrial plication, displacement of cardiac structures accounting for accidental injury to the right coronary artery, and relatively refractory postoperative atrial arrhythmias.

To the best of our knowledge, such rapid progression of right atrial dilation has not been reported, but

https://doi.org/10.1017/S1047951102000860 Published online by Cambridge University Press

probably represents the extreme end of the spectrum. This may suggest that patients with this relatively rare diagnosis should be monitored closely for potentially rapid progression needing intervention.

The respiratory symptoms were best explained on the basis of proximal compression of the airways secondary to massive dilation of the right atrium, even though the right atrium is an anterior structure. We hypothesized that, within the rigid confines of the chest cavity during episodes of agitation and struggling, there was exaggeration of mild proximal obstruction. Increased intrapleural pressure generated to overcome proximal obstruction would result in dynamic collapse of the distal airways. The same pathophysiology would also increase right heart pressures, and cause right-to-left shunting across the atrial septum, which would manifest clinically as cyanosis. The fluoroscopic findings collaborated these clinical inferences. For this reason, we strongly believed that surgery to reduce the size of the right atrium was indicated as the first intervention. The marked bulging of the right atrium seen on sternotomy, and the posterior displacement of the aorta found intraoperatively, provided anatomic evidence to support our hypothesis that the right atrium was responsible for compression of the airways. Postoperative resolution of the symptoms and virtually normal dynamic fluoroscopy obtained at follow-up within 2 weeks of the operation, then provided physiologic data to support the hypothesis, and ruled out underlying additional pathology in the tracheobronchial tree.

Since the diagnosis is relatively rare, it is important for surgeons to recognize that massive dilation may distort normal anatomy, and there is a need to define various anatomical landmarks to avoid injury to major structures.

There have also been suggestions that atrial reduction may be indicated to prevent further dilation.[3] Atrial arrhythmias may be a significant post-operative problem, as seen in our patient, and this should be taken into consideration when recommending this type of surgery in asymptomatic patients.

Acknowledgement

The authors would like to thank Dr Sameer Shah, Pediatric Critical Care, for his insights into the case.

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