

## Case reports

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# Giant blood cyst of tricuspid valve *Successful excision in an infant*

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*A case of a giant blood-filled cyst of the tricuspid valve is described in a 4-month-old infant. The cyst caused obstruction of the right ventricular inflow and outflow tracts and a right-to-left shunt; it was successfully removed at open heart surgery. Pitfalls in differential diagnosis and the pathogenesis are discussed.*

Cardiac tumours in infancy are exceedingly rare, particularly primary ones (Nadas and Ellison, 1968; Van der Hauwaert, 1971). Among these, to our knowledge, no case of symptomatic giant blood-filled cyst of the heart valves has been reported.

Small, pin-point cysts of the atrioventricular valves, usually multiple and located on the atrial surface of the leaflets, are a frequent finding at necropsy in infants under 6 months of age (Boyd, 1949).

The following case report is concerned with a 4-month-old infant who was successfully operated upon in our hospital for removal of a giant cyst of the tricuspid valve.

### Case report

A 4-month-old baby girl, weighing 4.9 kg, was referred to the Cardiology Department by her family doctor with a generic diagnosis of congenital heart disease. At birth, she had difficulty in breathing. At 1 month of age, an increase in heart size associated with cyanosis at rest led to the referral diagnosis and she was treated with digitalis.

At admission she was a well-nourished and developed baby with no signs of distress. However, cyanosis of the finger nails and visible mucosae was present. Heart rate was 150 and regular; systolic blood pressure was 95 mmHg (12.6 kPa).

Examination of the heart revealed a right ventricular lift without thrills. The first heart sound at the apex was loud and widely split, the second was single and weak. There was an  $S_4$  gallop and, at times, an  $S_3 + S_4$

slightly louder at the apex. A grade 2/6 short systolic regurgitant murmur was audible at the apex and at the left of the sternum. The liver was barely palpable below the right costal margin.

The haemoglobin level was 13.5 g/dl, haematocrit 52 per cent, and white blood count 11 200/mm<sup>3</sup>.

Chest x-ray examination showed moderate cardiomegaly, with reduced pulmonary vascular markings. The electrocardiogram showed sinus tachycardia, the QRS axis at +80 degrees, and right atrial and ventricular hypertrophy.

During cardiac catheterization, blood oxygen saturations were determined in the right and left heart chambers, and revealed obvious desaturation in the right atrium and ventricle, and a right-to-left shunt through a patent foramen ovale (left atrial saturation 42%). Right atrial pressure was 8 mmHg (1.1 kPa), left atrial pressure 7 mmHg (0.9 kPa); right ventricular pressure was reduced to 11/0–2 mmHg (1.5/0–0.3 kPa). The catheter could not be passed into the pulmonary artery.

On cineangiography, the contrast media, injected into the right ventricle, showed the cavity to be partially occupied by a bean-like mass, freely moving up and down through the tricuspid valve, probably connected to its septal leaflet (Fig. a). Regurgitant flow into the right atrium promptly visualized the left heart cavities and the aorta. The mass was clearly visible when contrast medium was injected directly into the right atrium (Fig. b). Forward flow slowly passed through an apparently rigid pulmonary valve. The final diagnosis was a right atrial mass. Surgical intervention was recommended.

The heart was explored through a median sternotomy and normothermic total cardiopulmonary bypass. The

right atrium was filled by a bluish, smooth mass extending through the tricuspid valve into the right ventricle and attached to the septal leaflet by a thin, 5 mm long pedicle. The mass was removed with its valvular insertion, which was repaired by suture.

Macroscopical examination of the tumour disclosed a unilocular cyst, 3 × 2 cm, filled with blood and with fibrin layers on the inner surface. The cystic wall was 1 mm thick and its histological examination showed oedematous connective tissue rich in fibroblasts and collagenous bands and scarce in ground substance. No elastic fibres were shown by Weigert staining. Both the inner and outer sides of the wall were lined with a thin endothelium. The cyst pedicle had a central lumen through which the cavity communicated with the right ventricle.

The patient's recovery was uneventful. At follow-up after 2 years, the child was active and developing normally; her heart size was much reduced and the electrocardiogram was within normal limits.

### Discussion

Despite an exhaustive search, no reports of symptomatic giant cyst of the tricuspid valve with diagnosis during infancy and successful removal were found, even though the high postmortem incidence of small blood cysts of the heart valves in infants is well documented. There are a few reports of large cysts of heart valves in children of school age and in young adults (Liese, Brainard, and Goto, 1963; Cumming and Ferguson, 1965; Sakakibara *et al.*, 1967; Leatherman *et al.*, 1968). These cysts, which caused more or less severe symptoms of valvular stenosis, were correctly diagnosed and surgically removed from the pulmonary valve in three instances (Liese *et al.*, 1963; Cumming and Ferguson, 1965; Sakakibara *et al.*, 1967), and the mitral valve in one (Leatherman *et al.*, 1968).

In our case, the cyst cavity was unilocular, filled with blood, and lined with endothelium. It communicated through a fine pedicle, implanted on the atrial side of septal leaflet of the tricuspid valve, with the cavity of the right ventricle. This finding supports Boyd's pathogenetic theory that these cysts are formed by blood pressed into crevices on the ventricular surface of the valves, with subsequent sealing off of the portal of entry (Boyd, 1949). However, the size of the mass we observed and the absence of elastic fibres in its walls suggest that the cyst originated more like a diverticulum of the septal tricuspid leaflet, most probably as a result of a localized structural defect. On the other hand, the existence of a multiloculated haematic cyst with fibroelastic elements in the stroma (Cumming and Ferguson, 1965) and the finding of an intracardiac penduculated lymphangioma (Forel, 1919) lead us

to speculate about a hamartomatous pathogenesis in these cases.

Another interesting point in our patient was the preoperative differential diagnosis. Some of the more

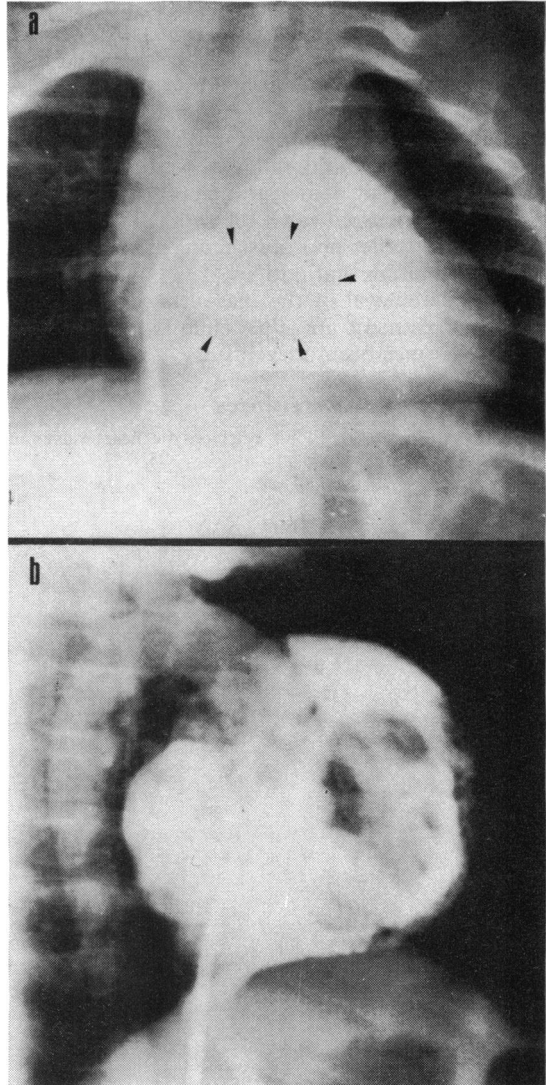


FIG. a) Selective right ventricular angiogram (AP projection, early phase), showing a bean-like filling defect (between arrows), partially occupying the right atrial and ventricular cavities. b) Selective right atrial injection (lateral projection). Late diastole: the mass extends also to the proximal outflow portion of the right ventricle. A right-to-left shunt is present at the atrial level which visualized the left heart structures.

impressive clinical findings, such as the cyanosis, early heart failure, and triple and sometimes quadruple rhythm associated with a systolic murmur, would have led us to suspect an Ebstein anomaly or a pedunculated intracavitary tumour. However, the relatively small size of the heart and electrocardiographic pattern of right ventricular hypertrophy were against the first, while the extreme rarity of intracavitary tumours in children under 4 years of age and the absence of conduction disturbances excluded the second possibility. The patient's symptoms and clinical findings were clarified by angiocardiographic demonstration of a mass moving through the tricuspid valve, following the excursions of its septal leaflet and obstructing both the right ventricular inflow and outflow.

Surgical removal of the mass has proved to be effective treatment for this child whose clinical course has since been uneventful.

#### References

- Boyd, T. A. B. (1949). Blood cysts on the heart valves of infant. *American Journal of Pathology*, **25**, 757.
- Cumming, G. R., and Ferguson, C. C. (1965). An elusive tumor of the pulmonary valve associated with a coronary arteriovenous fistula. *Journal of Thoracic and Cardiovascular Surgery*, **50**, 715.
- Forel, F. C. (1919). Two cases of primary neoplasm of the cardiac valves. *International Clinics*, 29th Series, **4**, 147.
- Leatherman, L., Leachman, R. D., Hallman, G. L., and Cooley, D. A. (1968). Cyst of the mitral valve. *American Journal of Cardiology*, **21**, 428.
- Liese, G. J., Brainard, S. C., and Goto, U. (1963). Giant blood cyst of the pulmonary valve. *New England Journal of Medicine*, **269**, 465.
- Nadas, A. S., and Ellison, R. C. (1968). Cardiac tumors in infancy. *American Journal of Cardiology*, **21**, 363.
- Sakakibara, S., Katsuhara, K., Iida, Y., and Nishida, H. (1967). Pulmonary subvalvular tumor. *Diseases of the Chest*, **51**, 637.
- Van der Hauwaert, L. G. (1971). Cardiac tumours in infancy and childhood. *British Heart Journal*, **33**, 125.

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