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Short report

Central muscular ventricular septal defect and coarctation of the aorta

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Many patients with coarctation of the aorta who present with severe heart failure in the first months of life have an associated ventricular septal defect. The need for surgical treatment of the coarctation at an early age has been widely accepted, but the appropriate treatment of the ventricular septal defect remains to be established.

During the five years 1980–4, 14 patients were seen with the diagnosis of coarctation and ventricular septal defect; their ages ranged from one week to five months. Five ventricular septal defects were of the central muscular type, two being small and the other three very large. We report the three patients with coarctation and a large central muscular ventricular septal defect to illustrate the natural history of this type of defect.

Case reports

CASE 1

A three week old boy was admitted to the hospital with signs of severe congestive heart failure. On physical examination palpation revealed intensive heart pulsations, hepatomegaly, and intermittently absent femoral pulses; auscultation disclosed loud heart sounds and a grade II ejection systolic murmur. The chest radiograph showed an enlarged heart (cardiothoracic ratio 0·75) and pulmonary plethora. At cardiac catheterisation the aortic isthmus showed tubular hypoplasia, the ductus was patent, and there was a large central muscular ventricular septal defect with equal right and left heart pressures. During subsequent operation the ductus was closed, the aorta was repaired by a subclavian flap, and banding of the pulmonary artery was performed.

At the age of 11 months a second catheterisation showed an intact ventricular septum; the aortic valve proved to be bicuspid, without appreciable stenosis. The band was removed from the pulmonary artery and the child is now in perfect condition.

CASE 2

A critically ill boy was admitted at the age of 3 weeks, severely decompensated and in circulatory shock. Two dimensional echocardiography established the presence of a localised coarctation and a large central muscular ventricular septal defect (fig 1). After artificial ventilation and prostaglandin E_1 infusion his condition improved dramatically. At operation the ductus was closed and the coarctation resected. Cardiac catheterisation at the age of 2 months, because of persistent heart failure, again showed a large defect with slightly pulmonary hypertension. From then on the heart failure diminished and the baby started to grow. Echocardiography at the age of 8 months showed an intact ventricular septum.

CASE 3

This boy was admitted to hospital at 2 weeks of age with all the signs of severe congestive heart failure. Cardiac catheterisation showed a large central muscular ventricular septal defect with severe pulmonary hypertension and a long, narrow aortic isthmus without localised coarctation. Medical treatment resulted in gradual improvement of the condition.

By the age of 6 months the signs of heart failure had disappeared. Repeat catheterisation showed again a large defect (fig 2); the diameter of the aortic isthmus, however, had become normal, as had the pulmonary artery pressure. At the age of 1 year an early systolic murmur of a small septal

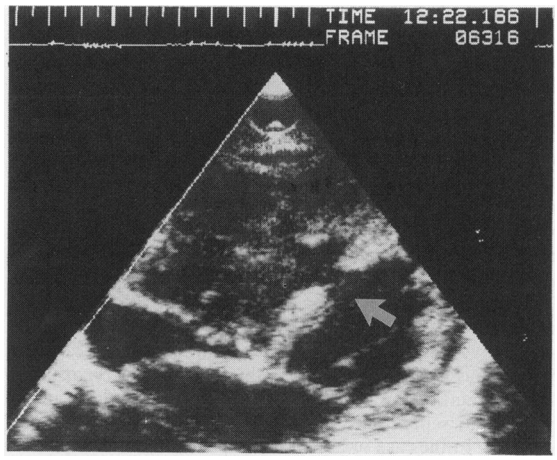

Fig 1 *Echocardiogram showing central muscular ventricular septal defect (case 2).*

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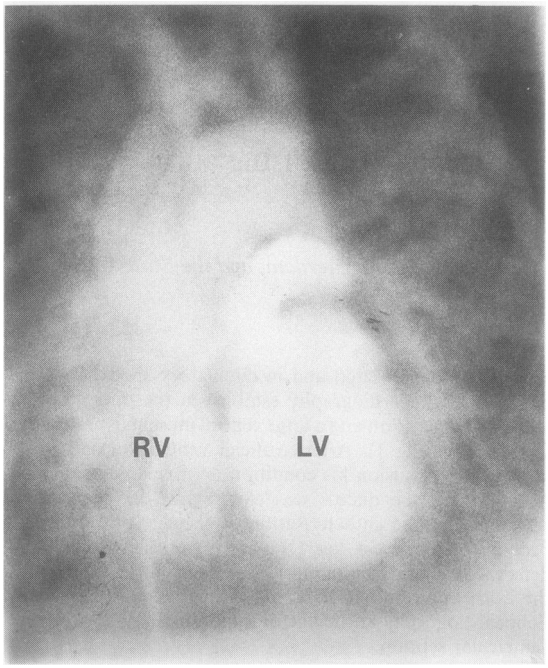

Fig 2 *Angiocardiogram showing central muscular ventricular septal defect (case 3). RV—right ventricle; LV—left ventricle.*

defect was still audible; chest radiography showed normal heart size and echocardiography showed the defect to be closed.

Discussion

The incidence of spontaneous closure of isolated ventricular septal defects may be as high as 50% within the first 10 years of life; small defects are especially likely to close, as are muscular defects and perimembranous defects extending into the trabecular and inlet septum.[1 2] The mechanisms responsible for closure are supposed to be growth of the muscular septum and adherence of the tricuspid valve leaflets. Clearly therefore in isolated defects the type and localisation of the defect are among the factors determining the prognosis. In infants with coarctation of the aorta a complicating ventricular septal defect is present in about half of the patients.[3 4] The increased afterload in the systemic circulation in patients with coarctation increases the magnitude of the left to right shunt through the septal defect, thereby magnifying the haemodynamic importance of the defect.[5]

The coexistence of an increased afterload due to the coarctation and thus an increased shunt results in the severe congestive failure found in these ill infants. The treatment policy that is generally accepted at present is to repair only the coarctation at the initial operation, and to close the ventricular septal defect subsequently as the clinical course dictates. In the case of heart failure persisting after repair of the coarctation, it must be decided whether to treat the patient medically or surgically. In this context it is important to know that isolated ventricular septal defects complicating coarctation may decrease in size or even close,

Remarkably little information has been published, however, regarding the type and localisation of defects associated with coarctation,[3-5] factors that influence the possibility of spontaneous closure. In 25 hearts with coarctation Anderson *et al*[6] found 24 perimembranous defects, partially closed by tricuspid valve tissue, and one muscular outlet defect. The authors concluded that the clinical implications of these findings remain to be evaluated.

Recently we reported the remarkably large incidence of aortic arch anomalies in hearts with a central muscular ventricular septal defect.[7] Among 25 heart specimens (including three with the architecture of a spontaneously closed defect) only six (24%) had an aortic arch of normal size and configuration.

The present clinical experience with three patients illustrates the concurrence of coarctation with a central muscular ventricular septal defect, and highlights the possibility of a favourable natural history with spontaneous closure. Normalisation of a hypoplastic aortic isthmus, as in case 3, is rare in our experience, thus repair of the coarctation will be necessary in practically all patients. After the intervention we advise medical management up to the age of 1 year, especially in patients with a central muscular defect; this will give spontaneous closure a fair chance, and thus avoid open heart surgery which may offer technical difficulties (necessitating a left ventriculotomy) and the risk of surgical heart block. After the age of 1 year large defects should be closed to prevent changes in pulmonary vascular resistance.

From the present experience of spontaneously closing large central muscular ventricular septal defects, we consider that there is no indication for pulmonary artery banding in these patients.

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