Case reports

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Double outlet right ventricle with subvalvular aortic stenosis¹

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SUMMARY A case of double outlet right ventricle had progressive muscular subvalvular aortic stenosis unrelated to the ventricular septal defect. Ventricular systolic pressures were identical and higher than aortic, and the gradient was within the right ventricle. Selective angiocardiology showed a hypertrophied subaortic conus obstructing the right ventricular outlet. Serial haemodynamic and angiographic studies revealed progression of the subaortic stenosis which may have been related to an earlier pulmonary artery banding operation. Distal conal hypertrophy is postulated as the cause of the obstruction.

The anatomical, pathophysiological, medical, and surgical aspects of double outlet right ventricle are well known. Edwards *et al.* (1952), Lauer *et al.* (1960), Neufeld *et al.* (1961a, b), Cheng (1962), Serratto *et al.* (1967), Mason *et al.* (1969), Lavoie *et al.* (1971), Rao and Sissman (1971), Megarity *et al.* (1972), Pellegrino *et al.* (1973), Goor and Lillehei (1975), and Cameron *et al.* (1976) have described complex variants of this lesion including associated mitral stenosis or atresia, pulmonary stenosis or atresia, coarctation of the aorta, persistent ductus arteriosus, and left ventricular obstruction from a restrictive ventricular septal defect. Left ventricular obstruction caused by subvalvular aortic stenosis has been cited by Mason *et al.* (1969) and by Spidaromont *et al.* (1976). We present haemodynamic and angiographic evidence of a patient with double outlet right ventricle with progressive muscular subaortic stenosis.

Case report

This 14-month-old girl weighed 2410 g at birth after a 37-week uncomplicated gestation. Within 2 hours of age she was found to have jejunal atresia. Shortly thereafter she developed congestive heart failure. Digoxin and frusemide improved her cardiac status and she successfully underwent abdominal surgery at 2 days of age. Cardiovascular examination disclosed a tachypnoeic, acyanotic infant whose

lungs were clear to auscultation. The arterial pulses were normal. The praecordium was hyperactive and the point of maximum impulse was diffuse along the left sternal border. The second heart sound was split and there was a loud third sound at the apex. A grade 3/6 long systolic ejection murmur was heard along the left sternal border. A mid-diastolic rumble was present at the apex. The liver was palpable 3 cm below the right costal margin. The electrocardiogram and vectorcardiogram showed right ventricular hypertrophy and the chest x-ray film showed cardiomegaly, with increased vascularity.

At 2 weeks of age, cardiac catheterisation showed a torrential pulmonary blood flow, and equal systolic pulmonary artery, aortic, and ventricular pressures (Table). Cineangiography showed a double outlet right ventricle with a large ventricular septal defect and suggested muscular subaortic stenosis (Figs 1 and 2). At 3 weeks of age, she underwent pulmonary artery banding and went home at 5 weeks of age much improved.

At 14 months, she presented as a small, comfortable, acyanotic toddler. The praecordium was slightly hyperactive and there was a thrill at the upper left and right sternal borders. The first heart sound was normal. The second heart sound was well split. There was a long harsh systolic ejection murmur maximal at the mid left sternal border radiating to the upper right and left sternal borders. A short mid-diastolic rumble was present at the apex. A sharp liver edge was palpable 2 cm

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Table Cardiac catheterisation data

Site	Pressure (mmHg)	O ₂ sat. (%)	Pressure (mmHg)	O ₂ sat. (%)
Superior vena cava		58		62
Right atrium		$\bar{m} = 2$ 71†	$\bar{m} = 5$	56
Right ventricle	77/4	89	164/12	
Main pulmonary artery	78/25	$\bar{m} = 51$ 87	*Prox. 123/12	$\bar{m} = 66$
			†Dist. 24/10	$\bar{m} = 14$ 82
Left atrium		$\bar{m} = 8$		$\bar{m} = 9.5$
Aorta	72/30	$\bar{m} = 51$ 89	80/45	$\bar{m} = 63$ 93
Left ventricle	70/7	89	164/14	93
Pulmonary blood flow (l/min per m ²)	8.9		8.7	
Systemic blood flow (l/min per m ²)	2.0		4.1	
Pulmonary to systemic blood flow	4.5:1		2.4:1	
Pulmonary vascular resistance (mmHg/l per min per m ²)	4.8		0.6	
Systemic vascular resistance (mmHg/l per min per m ²)	24.5		15.9	

*Proximal to band.

†Distal to band.

‡Left-to-right shunt at the atrial level, probably through a 'stretched' patent foramen ovale.

below the right costal margin. The electrocardiogram and vectorcardiogram showed combined ventricular hypertrophy. There was cardiomegaly and increased pulmonary vascularity on chest x-ray film. At repeat study there was moderately increased pulmonary blood flow (pulmonary: systemic flow ratio = 2.4), and manometry showed equal systolic ventricular pressures exceeding aortic pressure, and proximal main pulmonary arterial pressure exceeding aortic and distal pul-

monary arterial pressures. In addition, there was a 41 mmHg subpulmonary gradient. Cineangiography showed a double outlet right ventricle, adequate pulmonary artery band, subvalvular pulmonary stenosis and severe subaortic stenosis, and a large ventricular septal defect (Table; Fig. 3).

Discussion

Cases of double outlet right ventricle with left

Fig. 1 Selective right ventricular angiogram in the anteroposterior view shows the right ventricular origin of the aorta from a narrowed subaortic conus (arrow) to the right of the pulmonary artery. Abbreviations: AO, aorta, PA, pulmonary artery, RV, right ventricle.

Fig. 2 Left ventricular cineangiogram in left anterior oblique projection, showing the large ventricular septal defect (D), associated with double outlet right ventricle. The arrow shows the mitral valve, indicating the absence of aortic-mitral valve continuity. Abbreviations: AO, aorta, PA, pulmonary artery, RV, right ventricle, LV, left ventricle.

Fig. 3 Selective left ventricular cineangiograms in the anteroposterior projection obtained during the first (A) and second (B) cardiac catheterisations, showing the progression of the muscular subaortic stenosis (left-sided arrows). The right-sided arrow in (B) shows the moderately severe subpulmonary stenosis. Abbreviations: AO, aorta, LV, left ventricle, PA, pulmonary artery.

ventricular obstruction from a restrictive ventricular septal defect, reported by Edwards *et al.* (1952), Lauer *et al.* (1960), Neufeld *et al.* (1961a, b), Cheng (1962), Serratto *et al.* (1967), Mason *et al.* (1969), Lavoie *et al.* (1971), Rao and Sissman (1971), Megarity *et al.* (1972), and Pellegrino *et al.* (1973), have shown the systolic pressure of the left ventricle to be higher than that of the right ventricle, and equal systolic pressures in the right ventricle and in the aorta.

In 2 of 88 cases reported by Spidaromont *et al.* (1976) subaortic stenosis was suggested by a systolic pressure gradient between the right ventricle and aorta. There were no left ventricular pressures or angiographic data available in their cases. They suggested that the obstruction was the result of a hypertrophied parietal band. Mason *et al.* (1969) described angiographic and intraoperative findings of a double outlet right ventricle with aortic obstruction from a subaortic membrane. However, there was no gradient between the aorta and right ventricle.

The present case of double outlet right ventricle is unique as equal ventricular systolic pressures, a progressive subaortic systolic gradient, and angiographic findings of progressive muscular subaortic obstruction are shown. At 2 weeks of age there was no gradient between right ventricle and aorta, though angiographically there was evidence of obstruction. The absence of a gradient may have been related to the diminished systemic flow index.

At 14 months, she had grown and the systemic flow index increased. A gradient of 78 mm was then found between the inflow and the subaortic regions of the right ventricle and the subaortic obstruction was more severe angiographically.

According to Griep *et al.* (1974), subpulmonary stenosis is known to occur after a pulmonary artery band caused by hypertrophy of the subpulmonary distal conal septum. In double outlet right ventricle the distal conal septum lies beneath both great arteries. We suggest that hypertrophy of the subpulmonary component may have displaced the subaortic portion to the right, and that displacement and hypertrophy of the subaortic conus has led to subaortic obstruction in our case.

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