CASE REPORT

FAMILIAL PATENT DUCTUS ARTERIOSUS

BY

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Patent ductus arteriosus is one of the commoner congenital cardiac anomalies that occur in more than one member of a family. Carleton *et al.* (1958) found 41 families, reported between 1941 and 1958, with two or more members suffering from the same congenital heart disease: in 17 instances, the lesion was a persistent ductus. Kjaerjard (1946) and Joyce and O'Toole (1954) each reported three siblings affected and patent ductus has been found in two generations (Walker, 1951; Record and McKeown, 1953; Starer, 1953; Nadas, 1957). Walker and Ellis (1940–1) described a family in which the father and 4 of his 8 children were affected, and Taussig (1947) mentioned a family where the father, two of his three children, and one of his two grandchildren all showed unmistakable evidence of a patent ductus.

This case is briefly described because the family history helped to make the correct clinical diagnosis in infancy before the development of a typical continuous murmur.

Case History

This girl was the first child of Irish parents, born after a normal pregnancy lasting 42 weeks. The delivery was normal and her birth weight was 6 pounds 8 ounces. On the fifth day of life the infant vomited and had slight central cyanosis. A loud systolic murmur was heard over the base of the heart.

At the age of four months she was admitted to Charing Cross Hospital because she was breathless after bathing and feeding. She now weighed 13 pounds 13 ounces and was breathless at rest with intercostal recession and a respiratory rate of 64 a minute. The liver was palpable 4 cm. below the costal margin and the spleen was just palpable. There were no râles in the chest and no peripheral œdema. The pulse was 128 a minute and was of increased volume in both the arms and legs. There was a harsh crackling pansystolic murmur with a crescendo in late systole. This murmur, which was accompanied by a thrill, was heard best at the third left interspace but radiated widely and was audible in the interscapular region. The second heart sound was loud but not abnormally split. An X-ray showed a large heart due mainly to left ventricular enlargement, a dilated pulmonary artery, and pulmonary plethora. The electrocardiogram showed the pattern of left ventricular hypertrophy.

The cardiac failure was treated with digitalis and there was dramatic improvement. At the age of 5 months, an early diastolic diminuendo murmur was also heard in the third left interspace. At 6 months of age a classical machinery (Gibson) murmur was heard.

Family History

The father of this patient and two of the father's sisters have all had operations for patent ductus arteriosus; another sister died in infancy with whooping cough, and there was one stillbirth. The paternal grandmother died, aged 48, with dropsy, which was thought to be due to mitral stenosis but no autopsy was performed. The paternal grandfather had been previously married; 5 of the 6 children of his first marriage had no heart disease, but the sixth died at the age of 37 of a strained heart muscle after serving in the Irish Army.

Comment

At the age of four months when only a systolic murmur was audible, the diagnosis of patent ductus arteriosus was suspected because of the increased pulse pressure, the left ventricular hypertrophy, the character of the murmur, and the family history. Dammann and Sell (1952) however, found an increased pulse pressure in subjects who were eventually proved to have a ventricular septal defect. Ziegler (1952) observed left ventricular hypertrophy in 4 of 5 infants under the age of six months with patent ductus arteriosus, but it may occur in ventricular septal defect also (Keith *et al.*, 1958). The murmur was pansystolic with a crescendo in late systole and maximal in the third left intercostal space: it was similar to the ductus murmur detected by Burnard (1958) in some newborn infants. The crackling quality of the systolic murmur due to a patent ductus in infancy has been emphasized by Keith *et al.* (1958) and the murmur in the infant described here was thought to have this crackling quality before its significance was known. A harsh pansystolic murmur maximal in the third left interspace and accompanied by a thrill is often heard in patients with ventricular septal defect.

Although at four months this patient had many of the features of a persistent ductus in infancy observed by Zeigler (1952), the differentiation from ventricular septal defect was made mainly on the character of the murmur and the family history. The family history was particularly striking in this child for her father and two of his sisters had each had an operation for patent ductus. This diagnosis now appears to be correct but the family history may be misleading, as Campbell (1949) has described a woman with patent ductus whose daughter had coarctation of the aorta. Also Record and McKeown (1953) found that 7 of 150 fraternities containing a patent ductus had other siblings with congenital heart disease but only one of these seven was another case of patent ductus.

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