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CASE REPORTS

Isolated Right Ventricular Hypoplasia with Atrial Septal Defect

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ISOLATED right ventricular hypoplasia with atrial septal defect is a rare congenital malformation of the heart; only seven authentic cases have been reported in the world literature which has been reviewed recently.[1] In the latter series the condition existed in three adults of the same family; otherwise all reported cases have been in cyanotic infants or children. Underdevelopment or malformation of the tricuspid or pulmonic valves may or may not be present, but a patent foramen ovale or atrial septal defect is always an accompanying anomaly.

Because of the rarity of this lesion and because only one such patient so far has been operated upon, and in that instance with but limited success, the authors considered that the following report of data concerning the diagnosis and treatment of a patient recently studied was worthy of recording.

A 6-year-old girl was seen for the first time at the Cardiac Clinic of the Kingston General Hospital in November 1961. At 18 months of age she had been seen in Vancouver, and at that time the suggested diagnosis was either a common ventricle or a truncus arteriosus. The original impression, on examination in the outpatient department, was that she had a transposition of the great vessels. She was admitted to hospital for further examination.

On examination she was a small and lively girl but appeared slightly mentally retarded. She had 3+ cyanosis and digital clubbing. There was no increase in jugular venous pulsation. The cardiac apex beat was palpable 2 cm. outside the midclavicular line and there was a Grade 3 to 4 (of 6) systolic murmur maximal at the left sternal edge in the third interspace. An ejection click was audible at the cardiac apex and at the left sternal edge; this is clearly shown on the phonocardiogram (Fig. 1). The second sound was single. No diastolic murmur was detected, femoral pulses were present and of good quality, and the blood pressure was 105/70 mm. Hg.

The electrocardiogram showed sinus rhythm and an axis of +50. There was right atrial and left ventricular hypertrophy (Fig. 2). A radiograph of the chest showed a cardiothoracic ratio of 10:17 and the vascularity appeared diminished (Fig. 3).

It was felt, at this stage, that she did not have the tetralogy of Fallot or a simple transposition of the great vessels. The diagnosis of a truncus arteriosus was not in keeping with the radiographic findings, and the cardiogram would be unusual for a single ventricle. Tricuspid atresia seemed to be the most likely lesion in this case but the electrocardiographic axis was normal, which was against this diagnosis.

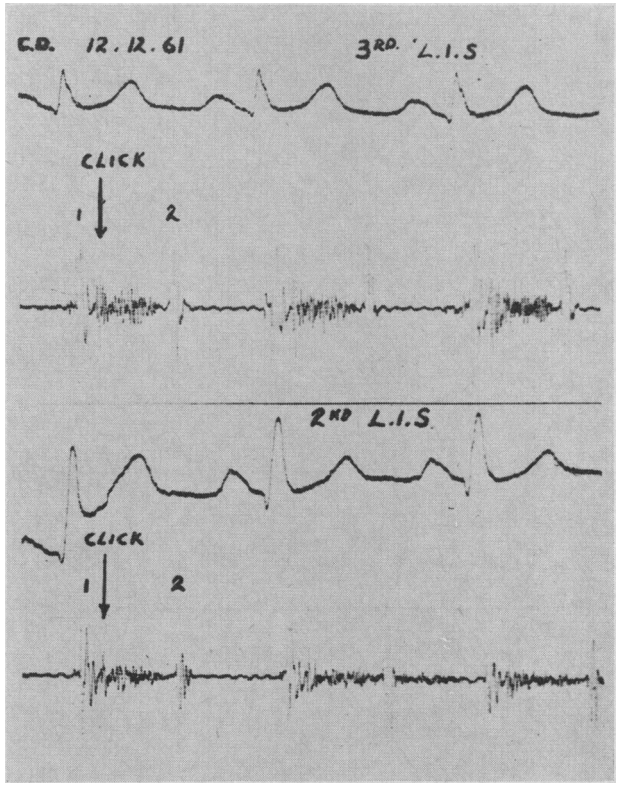

Fig. 1.—Phonocardiogram. Recording in second and third left interspaces shows systolic murmur with an ejection click.

On December 6, 1961, cardiac catheterization was carried out. The right ventricle was not entered but

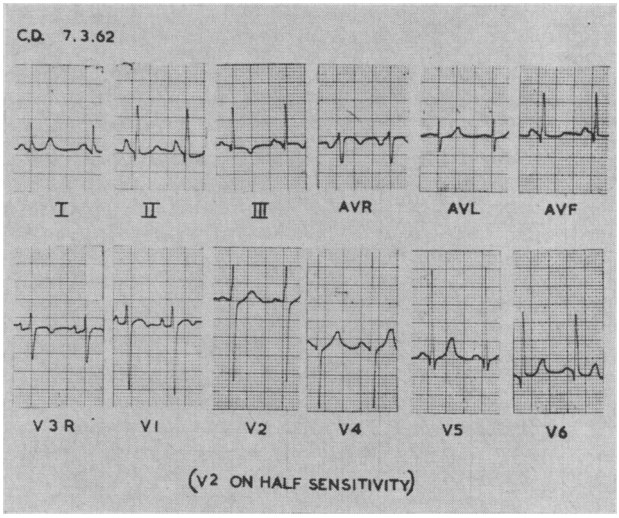

Fig. 2.—Electrocardiogram showing normal axis, right atrial and left ventricular hypertrophy.

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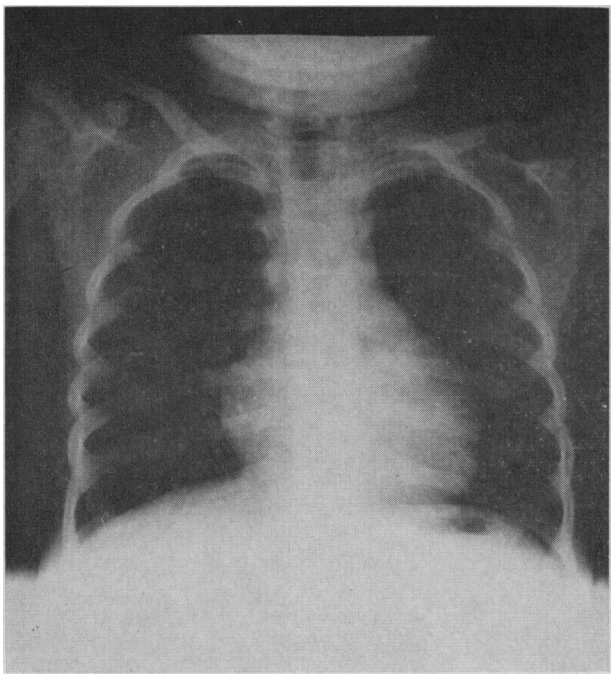

Fig. 3.—Chest radiograph. Cardiothoracic ratio of 54% with diminished vascularity.

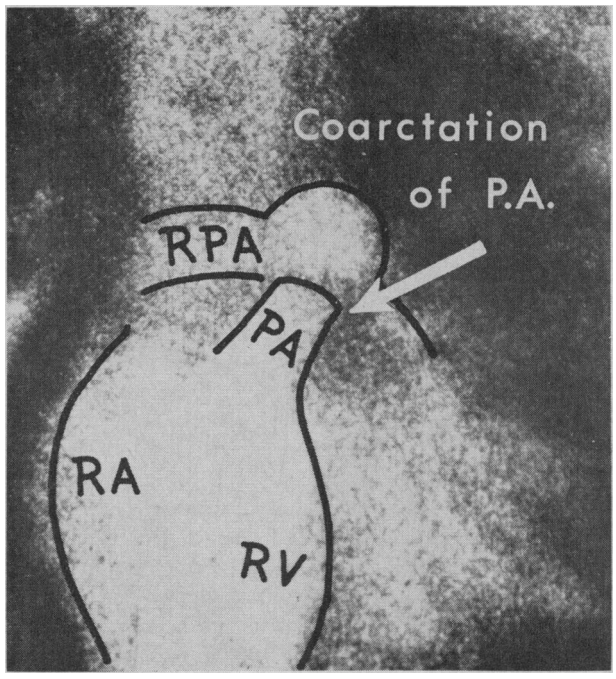

Fig. 4.—Angiocardiogram (taken from ciné film) showing small right ventricle and coarctation of the main pulmonary artery. Some of the dye has already shunted across the atrial septal defect and is filling the left ventricle.

the left ventricle was, and the pressure in this chamber was 100/0 mm. Hg, with an oxygen saturation of 70%. The catheter was withdrawn into the right atrium and angiocardiography was carried out. This demonstrated right-to-left shunting at the atrial level with rapid filling of a small right ventricle, and a coarctation of the main pulmonary artery. There was, however, clear filling of the right and left main pulmonary arteries beyond the area of stenosis (Fig. 4).

A diagnosis was made of hypoplastic right ventricle with an atrial septal defect and associated supravalvar pulmonary artery stenosis.

The patient was discharged to her home to have some dental work carried out and was readmitted to Kingston General Hospital on March 6, 1962. There had been no change in her clinical findings in the meantime. Her mother reported that just prior to readmission she had suffered "a blue spell" from which she had recovered fairly promptly. On March 12, 1962, a right thoracotomy was carried out. Hypoplasia of the right ventricle was confirmed and the right pulmonary artery was anastomosed to the superior vena cava, end-to-side. The superior vena cava was then ligated at its entrance into the right atrium, thus directing all the blood flow from the upper half of the body into the right lung, bypassing the heart. The postoperative course was uneventful, and it was obvious the day after her operation that her colour had improved. This improvement was confirmed on March 29 when a femoral artery sample showed the peripheral arterial saturation to be 83%. The patient was discharged to her home three weeks after operation.

The patient has been seen on several occasions since her discharge from hospital, most recently in January 1963. The clinical improvement was maintained; she was more active and had even been swimming. The mother commented on the improvement in her colour and also on the fact that her nails were less curved than they were preoperatively.

SUMMARY

A patient with the rare combination of isolated right ventricular hypoplasia with an associated atrial septal defect and pulmonary artery stenosis has been reported. Anastomosis of the right pulmonary artery to the superior vena cava improved the patient's clinical picture and exercise tolerance to almost normal levels.

The authors would like to thank Dr. A. M. Bryans, Professor of Pediatrics, for referring the patient, and Dr. S. L. Fransman of the Department of Radiology, for the angiography.

REFERENCE

1. SACKNER, M. A. *et al.*: *Circulation,* 24: 1388, 1961.

PAGES OUT OF THE PAST: FROM THE JOURNAL OF FIFTY YEARS AGO

THE DIAGNOSIS OF SMALLPOX

There was not one case, of the great number which I saw, in which the vesicles did not appear in crops, extending over many days, and in some cases they were covered from head to foot with vesicles and scabs at the same time, and yet not one vesicle was found to coalesce with another, and even in cases which were hideous to look at, in adults who had never been vaccinated, there was no secondary fever.

In an epidemic of smallpox in which the mortality was twenty-three per cent., a case of chicken-pox got into the smallpox hospital, where it contracted smallpox and died; yet Dr. Whitelaw tells us it is not a serious thing to mistake grippe or chicken-pox for smallpox.—Excerpt from letter to the Editor; H. H. McNally, *Canad. Med. Ass. J.*, 3: 210, 1913.