A CASE WITH THREE ATRIAL SEPTAL DEFECTS

BY

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This case is reported because of the unusual finding of the three common types of defect occurring in the one patient.

E.G., a girl aged 7 years 6 months, born in Holland of Dutch parents, was one of three children, the other two being in normal health. Her father died of "cyanotic heart disease" at the age of 25 years.

She had suffered from frequent cough and colds since infancy. She showed some limitation of exercise tolerance by shortness of breath on moderate exercise. She had always tired easily. Cyanosis had never been noticed by her parents or by medical attendants. An abnormal cardiac murmur had been noticed at 2½ years of age.

On examination she was a small elfin child of very fair complexion. She weighed 19 kg., was 112 cm. in height with an estimated surface area of 0.8 m.² She was not cyanosed, and there was no finger clubbing. Her chest was deep in the antero-posterior diameter, but there were no abnormal physical signs in the lungs. Blood pressure was 110/70 mm. Hg. The apex beat was in the fifth intercostal space, 25 cm. outside the mid-clavicular line, and was left ventricular in type, but the right ventricular contraction was easily palpable. The heart sounds revealed a normal first sound at the apex with a short mid-diastolic murmur. The second sound in the pulmonary area showed a "fixed split" with some accentuation of the pulmonary element. A moderate systolic murmur was heard all over the præcordium radiating towards the neck but was maximal along the left sternal border.

The electrocardiogram showed left axis deviation and "right bundle-branch block" pattern. The frontal plane vector formed a counter-clockwise loop with the main axis about -45°. The plain radiogram of the chest (Fig. 1) demonstrated some enlargement of the right ventricle and right atrium, with considerable enlargement of the main pulmonary artery and primary branches. The findings at cardiac catheterization (by courtesy of Dr. A. Venables, Royal Children's Hospital, Melbourne) are shown in Table I.

It was noted at the catheterization that "a right pulmonary vein was entered apparently directly from the right atrium."

The clinical picture and special investigations pointed to a diagnosis of left-to-right shunt at atrial level.

FIG. 1. X-ray showing enlargement of the heart, mainly of the right ventricle and right atrium, and of the pulmonary trunk and its primary branches.

TABLE I

Site	Saturation (%)	Pressure
S.V.C.	74	
R.A.	87 and 85	
R.V.	87	27/0
R.P.A.	87	20/3
L.A.	96	

The defect was thought to be an endocardial cushion defect Grade I (in Campbell and Missen's series), i.e. ostium primum defect with split mitral valve, and surgical treatment was planned, using the heart-lung bypass technique.

An operation was performed on November 8, 1961, with pentothal induction, nitrous oxide, oxygen, and halothane. Whole body perfusion was performed using a Melrose N.E.P. machine at a flow rate of 2.4 l./min./m.², and the perfusion time was 62 minutes at normal temperatures throughout.

Incision was bilateral thoracotomy along the lower borders of the fourth ribs with division of the sternum. Finger exploration via the right atrial appendage before bypass revealed the triple nature of the abnormality.

A summary of the macroscopic findings was as follows. The heart was considerably enlarged, and its configuration was consistent with a large interatrial shunt, and the abnormal veins from the right upper lobe were obvious on opening the chest. There was an obvious systolic thrill in the outflow tract of the right ventricle which was obliterated by the closing of the atrial defects. The superior vena cava and inferior vena cava were normal. There was no left superior vena cava. The right atrium was considerably enlarged, and the superior pulmonary vein on the right side entered the lower end of the superior vena cava opposite the sinus venosus defect which was approximately 1.2 cm. in diameter. There was a foramen ovale defect approximately 1.5 cm. in diameter, and a much larger "ostium primum" type defect with a low crescentic upper margin. There was no obvious deformity of the tricuspid valve, but there was a complete split of the anterior cusp of the mitral valve with no deformity of the valve ring (see Fig. 2). There was significant regurgitation

FIG. 2.—Drawing of the interior of the right side of the heart. It shows a sinus venosus defect above and to the left; a foramen ovale defect below this; and a much larger ostium primum defect (see text).

at the mitral valve before repair. The right ventricle was considerably enlarged, but its wall was not grossly hypertrophied. The pulmonary trunk was approximately twice the diameter of the aorta. Left atrium and left ventricle were not enlarged. There was no patent ductus arteriosus.

The sinus venosus defect was closed first by detaching the atrial septum posteriorly between the sinus venosus defect and foramen ovale defect and swinging the septal "flap" so created upwards across the sinus venosus defect and suturing it in position with a series of interrupted sutures which were tied on the external surface of the atrium. The split in the mitral valve was then sutured with a series of interrupted sutures. A single "teflon" felt patch was sutured into the enlarged foramen ovale defect and "ostium primum" defect, using a series of interrupted mattress sutures at the lower end and a continuous suture in two parts for the remainder of the margin. The bar of atrial septum between the foramen ovale and "ostium primum" defects was then fixed to the left side of the patch with three interrupted sutures. The atria fibrillated during the repair and this rhythm was still present at the end of the operation.

The post-operative course was as follows. The atrial fibrillation that developed at operation was treated by full digitalization at that time. A few hours after operation this had changed to a nodal rhythm which persisted for 12 days then reverted to sinus rhythm. She was discharged from hospital on the 23rd day after operation and her progress since has been satisfactory. When seen at the Cardiac Clinic 14 weeks after operation she was gaining weight, and her parents had noted a significant improvement in her exercise tolerance and general activity.

Comment

In a personal communication Mr. T. Holmes Sellors says that in his series of over 300 cases he has had several with two interatrial defects, but none with all three types. This series from the Royal Adelaide Hospital consists of 70 operations on 68 cases: the nature of the lesions and the modes of treatment are given in Table II.

TABLE II

Defects	Hypothermia	"Atrial Well"	Bypass	Total	Mortality
Foramen ovale type	9	25	16	50	Nil
Endocardial cushion defect					
(a) Grade I (Campbell and Missen)	1	0	8	9	Nil
(b) Grade II (Campbell and Missen)	0	0	1	1	Nil
(c) Grade III (Campbell and Missen)	0	0	1	1	1
Atrial septal defect with abnormal venous drainage					
(a) Sinus venosus	1	2	3	6	Nil
(b) Others	0	1	2	3	Nil
Total	11	28	31	70	1

The above series represents the total experience of this unit since the first case was attempted on December 11, 1958, but total extracorporeal circulation was not used before November 1960.

The present policy of this unit is to operate on uncomplicated foramen ovale defects in children up to their teens, using the "well" technique. All other cases with atrial septal defects (including foramen ovale defects in which the diagnosis is in any doubt) are managed with the assistance of extracorporeal circulation.

The difficulty of defining the exact nature, position, and size of the defect, using even the most refined methods of investigation, has led to a gradual change in policy which is continuing. All cases can be managed relatively easily with the aid of open heart technique in contrast to alternative methods, each of which has certain limitations in dealing with particular types of interatrial communication. It is probable that in the foreseeable future as the risks of extra-corporeal circulation are reduced all operations will be performed in this manner and that other methods will remain of historical interest only.

The author thanks Dr. J. L. Waddy for the electrocardiographic and phonocardiographic records, and Miss B. Beckett for the preparation of Fig. 2.

Reference

- Campbell, M., and Missen, G. A. K. (1957). Endocardial cushion defects: common atrio-ventricular canal and ostium primum. *Brit. Heart J.*, **19**, 403.