ENDOCARDIAL CUSHION DEFECTS IN INFANTS AND CHILDREN

TEN YEARS' SURGICAL EXPERIENCE

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For purposes of clarity the cardiac abnormalities referred to in this communication are grouped under the term "endocardial cushion defects" introduced by Watkins and Gross (1955) and simplified into 3 grades by Campbell and Missen (1957). The Table will help the reader identify the particular lesion if a different nomenclature is used. Failure of the endocardial cushions or masses of connective tissue to develop normally and fuse in the midline of the common atrio-ventricular canal may produce as many as 14 possible combinations of defects (Kiely *et al.*, 1958). It has been our experience that though no two lesions are exactly alike enough similarity exists to place each abnormality into one of the 3 grades described by Campbell and Missen (1957).

TABLE
NOMENCLATURE IN ENDOCARDIAL CUSHION DEFECTS

Reference		
Paul (1958)	Campbell and Missen (1957)	Wakai and Edwards (1958)
Persistent ostium primum with cleft mitral valve	Endocardial cushion defect, grade I	Persistent common atrio-ventricular canal, partial form
Persistent ostium primum with cleft mitral and tricuspid valves	Endocardial cushion defect, grade II	Persistent common atrio-ventricular canal, complete form
Persistent common atrio-ventricular canal	Endocardial cushion defect, grade III	Persistent common atrio-ventricular canal, complete form

The patients were referred for operation from the Department of Cardiology under the direction of Dr. John D. Keith at the Hospital for Sick Children, Toronto. Only those children in whom total correction was attempted are included. Excluded are infants with grade III defects where pulmonary artery banding was performed. Of a total of 84 cases, 56 were classified as grade I, 5 as grade II, and 23 as grade III. There were 39 male and 45 female patients, 5 of whom were mongoloids (all in grade III). One patient in grade I had an associated chondro-ectodermal dysplasia.

The majority of the patients with the grade I form were asymptomatic when first seen, but a careful history of each one revealed some degree of incapacity. Very slight to moderate dyspnoea on

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exertion was noted in 51 per cent; poor exercise tolerance in 48 per cent; but congestive heart failure in only 9 per cent of the patients. By contrast, of the children with the grade III form, 68 per cent had dyspnoea, 64 per cent had poor exercise tolerance, 55 per cent were in congestive heart failure, and 32 per cent had cyanosis at one time or another indicating a bidirectional shunt.

Auscultatory findings in these patients were not absolutely diagnostic. A pansystolic murmur at the apex was present in only 67 per cent of all the patients. A cleft mitral valve may be present with no apical murmur and normal left atrial pressure tracings. An ejection systolic murmur in the pulmonary area was a more common finding in all three groups and was often accompanied by a mid-diastolic flow murmur at the lower left sternal border. The clinical signs of moderate to severe pulmonary hypertension indicated by an accentuated second heart sound in the pulmonary area with narrow splitting was present in 27 per cent of the patients in the grade III form and in only 2 per cent of those in grade I.

The electrocardiographic finding of combined ventricular hypertrophy was predominant in all 3 grades; right atrial enlargement was present in 20 per cent. The most important and consistent finding in all these was the counterclockwise progression of the vectorial loop in the frontal plane. A left axis deviation of the QRS complex was present in all patients except two; however, both of these had counterclockwise loops.

Patients with the grade III form have much larger hearts than those in the grade I group; 45 per cent of the former had a cardiothoracic ratio over 61 per cent.

SURGICAL TECHNIQUE

All patients were operated upon using complete cardiopulmonary bypass and mild-to-moderate hypothermia. One patient, however, was done under hypothermia alone because of a mistaken pre-operative diagnosis of an atrial septal defect of the secundum type.

The approach was usually a sternal splitting incision; in some, particularly in girls, a right anterior thoracotomy without crossing the sternum was employed. A right atriotomy was performed close to and almost parallel to the interatrial groove. The right atrium was large in most cases and the operation could be done entirely through the right atrium. A careful assessment of the pathology was first undertaken before the repair was performed: in the presence of an associated secundum atrial septal defect the intervening septum between the ostium secundum and the ostium primum was divided. The clefts in the valves were closed using 4-0 interrupted silk sutures. The chordae tendineae were not divided even if they appeared to be foreshortened. If there was a wide cleft, a pericardial patch or a wedge of teflon was used to bridge the defect. In some cases, the valve ring was widely dilated and an annuloplasty was performed using mattress sutures at one or both commissures.

The septal defect was closed using a teflon patch with interrupted sutures at the base and continuous sutures at the free margin of the defect. Because of the proximity of the conduction mechanism, shallow but wide interrupted sutures were placed in this region. These sutures, interrupted and double-ended, were placed on the right side of the septum as an extra precaution to avoid heart block. Before final closure of the defect is accomplished, the mitral valve is rendered incompetent and the left atrium allowed to fill with blood, usually by ventilating the lungs. In grade III cases where the VSD is large it may be wise to place a patch from the right ventricular side, as was done in one of our most successful cases. Care must be taken to search for fenestrations at the base of the anterior leaflet of the mitral valve.

The right atriotomy is then closed and the patient taken off bypass. As soon as the systemic pressures are stabilized, a left atrial pressure is taken and if this shows significant insufficiency, the patient is placed back on bypass and an annuloplasty performed.

RESULTS OF SURGERY

Of the 84 patients operated upon, 26 died during the operation or immediately afterwards, an over-all mortality of 31 per cent. The highest mortality was in that group operated upon under the

FIG. 1.—Results of operation for endocardial cushion defects based on classification of Campbell and Missen (1957). See the Table.

age of 2 years. The mortality for the grade I type was 16.3 per cent. This included 2 patients who required re-operation for severe mitral incompetence and who died following the second operation. The mortality for the grade II group was 20 per cent and for grade III group, 73 per cent (see Fig. 1).

A persistent mitral regurgitant murmur was present in 27 patients or in 59 per cent of those who survived. Our cardiologists feel that in 24 of these the murmur is of little clinical significance. Some of these children, however, are on a maintenance dose of digitalis but recatheterization has not, as yet, been considered necessary. Eight patients were in intractable heart failure when operated upon and 5 of these patients died at operation. Pulmonary hypertension was present in 7, all with the grade III type, and 6 of these died following operation. Complete heart block was produced in 8 patients, and 3 of these died immediately after operation. The other 5 reverted to normal sinus rhythm at 3 hours, 8 days, 10 days, 13 days, and 2½ months after operation. Cerebral air embolism was produced in 3 patients, all of whom died after operation, showing evidence of brain damage. One patient died of an overwhelming *Staphylococcus aureus* septicaemia.

DISCUSSION

As experience was gained in the operative management of these defects certain technical aspects of the procedure assumed more importance. The grade I defects command more respect than we originally thought. The ostium primum defect should rarely be closed by direct suture since sutures placed deeply in the interventricular septum may produce catastrophic heart block. The use of a patch allows more superficial sutures under less tension above the bundle of His on the right side of the septum. In most cases suture of the cleft in the mitral valve produces a reasonably competent valve; however, persistent regurgitation is the rule rather than the exception. Indeed with the atrial septal defect closed mitral regurgitation may be severe enough to cause cardiac failure. Lengthening of the anterior leaflet, annuloplasty, and valve replacement at the initial operation may be necessary. As an example of the difficulties that can arise the following case is quoted.

A 7-year-old girl, nearly asymptomatic, was diagnosed as a grade I endocardial cushion defect and operated upon in the usual fashion. A cleft mitral valve was repaired and a tight chorda divided and the primum defect closed with a teflon felt patch. After bypass was discontinued the left atrial pressure was 15/7 with a

mean of 12 mm. Hg. We felt that this was an acceptable correction. Following operation she developed a hæmolytic anæmia which we felt was due to destruction of the blood as the regurgitant jet struck the teflon patch. She developed more severe mitral regurgitation and went into intractable cardiac failure. At a further operation two months after the first, at which time the left atrial pressure was 45/20 mm. Hg, the patch was removed and the mitral valve repair seemed satisfactory but the anterior leaflet appeared to balloon into the right atrium. An annuloplasty was performed at both commissures. The patch was repositioned and on coming off bypass the left atrial pressure was 60/30 mm. Hg. How we accomplished this is difficult to understand. The child was again placed on bypass and a Starr-Edwards valve replacement performed obtaining a left atrial pressure of 15/7 mm. Hg. She died three days later without regaining consciousness, presumably of cerebral air embolism. We believe that dividing the chorda was a technical error and we should have replaced it at the second operation.

If concern exists after whatever manœuvre, a small atrial septal defect should be created and proved life-saving in one of our patients. Children with grade II defects usually have a poorly developed septal leaflet of the tricuspid valve. Removal of this leaflet in the experimental animal causes very little incompetence of the tricuspid valve (Wait and Mustard, 1965). If the anterior and posterior leaflets appear adequate the valve may be competent. If doubt is present an annuloplasty inferiorly may produce a competent valve. On reviewing our experience of grade III defects it appears that in the first two years of life one should simply band the pulmonary artery. If total correction is contemplated one should be prepared to replace the valves. If intractable cardiac failure is present the prognosis is grave. The older child exhibiting bouts of cyanosis is probably inoperable due to pulmonary vascular disease. If the ventricular septal defect is large accurate closure through a right ventriculotomy after repair of the valves should be done: this was very successful in one of our patients. The over-all mortality was 31 per cent, 19 per cent of total in the first two years of life. Early in our experience heart block and air embolus proved fatal; proper technique should overcome these difficulties. We have used the nomenclature suggested by Campbell and Missen (1957), and though grade III carried a mortality of 70 per cent, 50 per cent of these deaths were in the first two years of life. Grade I lesions should command respect; if severe mitral insufficiency remains the child's condition is worsened by closure of the atrial septal defect.

SUMMARY

Eighty-four infants and children with endocardial cushion defects have been operated upon during the past 10 years at the Hospital for Sick Children, Toronto. Employing the classification of Campbell and Missen (1957), 56 were in grade I, 5 in grade II, and 23 in grade III. As we would expect, grade III defects were operated upon early in life with only one survivor in 13 cases. Banding of the pulmonary artery may offer some hope for these infants. Grade II defects are uncommon, with one death in 5 cases. Grade I defects are common in the age-group 2-15 years, and of 56 cases 6 died following operation, 2 of these following re-operation for severe mitral insufficiency. At least 60 per cent of all survivals have some degree of mitral incompetence. Grade I defects demand meticulous correction of mitral insufficiency and avoidance of heart block. In the older child a malformed mitral valve should be replaced if possible.

REFERENCES

- Campbell, M., and Missen, G. A. K. (1957). Endocardial cushion defects: common atrio-ventricular canal and ostium primum. *Brit. Heart J.*, **19**, 403.
- Kiely, B., Adams, P., Jr., Anderson, R. C., and Lester, R. G. (1958). The ostium primum syndrome. *A.M.A. J. Dis. Child.*, **96**, 381.
- Paul, M. H. (1958). Endocardial cushion defects: Persistent common atrioventricular canal and persistent ostium primum. *Pediat. Clin. N. Amer.*, **5**, 1011.
- Wait, J. H., and Mustard, W. T. (1965). Excision of the septal leaflet of the atrioventricular tricuspid valve. *J. thorac. cardiovasc. Surg.* In the press.
- Wakai, C. S., and Edwards, J. E. (1958). Pathologic study of persistent common atrioventricular canal. *Amer. Heart J.*, **56**, 779.
- Watkins, E., Jr., and Gross, R. E. (1955). Experiences with surgical repair of atrial septal defects. *J. thorac. Surg.*, **30**, 469.

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