nearer the true risk generally and the higher ones apply to particular epidemics.

Deafness is the greatest risk, then congenital heart disease, and then cataract: mental defect and microcephaly and several other malformations are lesser risks. All these may occur alone or quite often in combinations. Of the heart malformations persistent ductus is much the most common (58%); then comes ventricular septal defect (18%). In about 6% of these the two defects are combined, and in ordinary practice this is not a very common finding. Atrial septal defect, pulmonary valvular stenosis, and Fallot's tetralogy each occur in about 6% of cases. This distribution is quite unlike anything found under other conditions.

Numerically, maternal rubella during pregnancy is not one of the major causes of congenital heart disease, though it is one of the best-proved causes. It is responsible for something between 2 and 4% of all cases. At the Congress on Congenital Malformations in London in July, 1960, the proportion of *all these* caused by maternal rubella and *all other viral infections* was thought to be less than 10%.

Other viral infections, including mumps, measles, epidemic hepatitis, and poliomyelitis, can produce similar malformations. It seems unlikely that they often do so, though measles and even any minor infections during pregnancy need further investigation.

I am most grateful to Dr. Julia Bell for letting me have the details of the type of congenital heart disease in the reported cases she had collected.

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ENDOCARDIAL CUSHION DEFECTS

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The surgical implications involved in misdiagnosing an endocardial cushion defect as a simple patency of an ostium secundum may be very serious. This is because the former cannot always be corrected satisfactorily without the application of an extracorporeal circulation, whereas an ostium secundum can usually be closed in a shorter time with moderate hypothermia or by the use of an atrial well. It is therefore of great importance to be able to differentiate these two types of lesion with reasonable certainty pre-operatively, so that the surgeon may be enabled to select the appropriate technical methods to achieve repair with an open heart.

It is easy to make this differentiation in most cases on clinical and other grounds. However, there is a type of large ostium secundum, in which the lower margin of the defect may reach to within a few millimetres of the endocardial cushion area, which so closely resembles an endocardial cushion defect in all characteristics that we believe it to be virtually indistinguishable from the latter before cardiectomy. We have also encountered an endocardial cushion defect in which all the evidence pointed to a large ventricular septal defect with pulmonary hypertension. We think it will be useful to describe our experience with 10 proved cases of endocardial cushion defect and to compare them with the two cases of large ostium secundum which we incorrectly diagnosed as endocardial cushion defects before operation.

Our cases have all been seen in infancy or early childhood and have been fully investigated, with the exception of one infant who died at 2 months before cardiac catheterization was performed. The diagnosis in all has been verified by visual inspection at operation or necropsy.

Nature of the Endocardial Cushion Defect

The embryological failure which gives rise to the varying degrees of endocardial cushion defect is well known, and excellent descriptions are given by Kramer (1942), Wakai and Edwards (1956), and Campbell and Missen (1957). It is clear that failure of normal fusion of the dorsal and ventral endocardial cushions of the primitive atrio-ventricular canal can lead to a spectrum of lesions extending from persistence of the ostium primum with deformity of the atrio-ventricular valve cusps to the complete anomaly in which, the cushions having failed to approximate, there is a persistency of the atrio-ventricular canal with a functionally unilocular heart. Numerous variations of the complex have been described, and a review of these is given by Kiely *et al.* (1958) together with a tabulation of 14 possible combinations of the defects comprising the syndrome.

Ostium primum patency without atrio-ventricular valve deformity appears to be exceedingly rare, if it ever occurs. Wakai and Edwards (1958), in a recent review of 100 cases of endocardial cushion defect, could

find no authentic case. They excluded the case of Blount *et al.* (1956) as the published picture of the heart showed a defect too posterior in position to be, in their opinion, an ostium primum.

There is, unfortunately, a lack of uniformity in the nomenclature applied to these variants, and they have been described under such names as ostium primum defect, partial or complete persistence of the atrio-ventricular canal, atrio-ventricularis communis, and endocardial cushion defects. Since the septational and valvular defects stem from the basic disorder in growth and fusion of the endocardial cushions, it seems logical to us to apply the generic term "endocardial cushion defect" to all varieties of this syndrome as advocated by Campbell and Missen (1957) and developed by them from the usage of Watkins and Gross (1955). The subdivision of cases into three grades dependent on the extent of the abnormality is also a great help in clarifying description. Thus grade I includes cases of persistence of the ostium primum with splitting of the anterior cusp of the mitral valve, or the septal cusp of the tricuspid valve, or both; grade II cases with a persistent ostium primum, bifid atrio-ventricular valve cusps, and a high ventricular septal defect; and grade III those with persistence of the common atrio-ventricular canal.

Clinical Material

The 10 cases of endocardial cushion defect and two cases of large ostium secundum patency were found during a period of eight years (1952-9) in which a total of 248 new cases of congenital cardiovascular disease were investigated in the children's department at Westminster Hospital. Of these patients, 66 (over 25%) were under 1 year of age. In this series the incidence of endocardial cushion defects is 4%, but it cannot be inferred that the true incidence is so high, since our patients are a highly artificial selection with a predominance of incapacitated infants and young children. Keith *et al.* (1958) state that about 2% of all congenital heart anomalies in children up to 14 years are of this type. Bailey *et al.* (1954) and Watkins and Gross (1955) report a 10-15% incidence of endocardial cushion defect encountered during operations for atrial septal defects. Paul (1958) says that in his experience in Chicago "one endocardial cushion defect presents itself for each seven or eight ventricular septal defects."

Only two of our cases were mongols, although during the same period eight mongol children with congenital heart disease have been fully investigated. This, although admittedly a small number on which to place much emphasis, is at variance with some of the published experience; for example, Paul (1958) states that the mongol with congenital heart disease has a 30-50% possibility of having an endocardial cushion defect.

All the cases (10 endocardial cushion defects and two large ostium secundum patencies) presented in the first two years of life with symptoms indicative of a disabling heart lesion, with the exception of one child (Case 8) with a grade I cushion defect who was asymptomatic when successfully operated upon at 8 years (see Table I). In many of the children who survived infancy the symptoms have become less severe with the passage of time, a tendency noted by most observers in a high proportion of cases with a predominant left-to-right shunt. Three infants, however, died in the first year of life of heart failure (Case 1, 5 months; Case 2, 2 months; Case 9, 8 months).

Another child (Case 4), after a life of 5½ years characterized by frequent severe respiratory illnesses and almost complete cessation of growth after 4 to 5 months of age, died of combined pneumonia and congestive heart failure. His clinical signs and electrocardiogram showed the sequential changes of mounting pulmonary hypertension. This child had a grade I defect, Case 2 a grade II defect, and Cases 1 and 9 grade III defects. Cases 2 and 9 also had patent ductuses.

The presenting symptoms are similar to those of any other severe intracardiac defect. No one physical sign is pathognomonic of this condition. A loud, rasping holosystolic murmur maximal in the lower left parasternal area, but widely conducted over the whole praecordium and radiating into the axilla, was heard in seven of the cases of endocardial cushion defect and in both the cases of large ostium secundum patency. In many the murmur was accompanied by a thrill. It is extremely difficult, in our experience, to differentiate this murmur from that heard in many cases of ventricular septal defect, especially when it is present with a thrill. A low-pitched mid-diastolic murmur heard at, or just internal to, the apex was noted in five cases of endocardial cushion defect and in both cases of ostium secundum patency. The finding of this murmur in the two latter makes it difficult certainly to ascribe its presence in endocardial cushion defects to deformity of the mitral valve. It may possibly be due to the large flow of blood through the atrial defect or to relative tricuspid stenosis consequent on the left-to-right atrial shunt, since these are features common to both types of cases. A representative phonocardiogram showing the apical diastolic murmur is presented in Fig. 1.

FIG. 1.—Phonocardiogram from Case 6 with a grade I endocardial cushion defect showing the mid-diastolic apical murmur.

Chest Radiography

This appears to be of little value in discriminating between endocardial cushion defects and other lesions with free communication between the left and right heart chambers and a predominant left-to-right blood shunt. Both the cases with a large low-sited ostium secundum showed no distinguishing features from the

TABLE I.—*Clinical Details*

Case No.	Date of Birth	Infantile History	Major Symptoms	Cardiovascular Signs	Nature of Defect	Subsequent Progress
1	24/9/57	Full-term, normal pregnancy. Normal delivery. Birth weight 7 lb. 11 oz. (3,485 g.). Well until 6 weeks, when admitted to hospital with pneumonia. Congenital heart disease diagnosed. Transferred Westminster Hospital 2/12/57. Recurrent cyanotic attacks. Treated with digoxin and oxygen	Loss of weight. Breathlessness. Cyanotic attacks	Very large heart. Apex beat in 6th left intercostal space in anterior axillary line. Soft blowing systolic murmur maximal in pulmonary area	E.C.D. Grade III	Cardiac catheterization and selective angiocardiology 2/1/58. Died in congestive heart failure 1/2/58
2	25/3/56	Full-term, normal pregnancy. Normal delivery. Birth weight 7 lb. 10 oz. (3,460 g.). Occasional vomiting until 5 weeks, when cyanosis sometimes noticed with vomiting. Poor weight gain. By 8 weeks was severely cyanosed when vomiting. Admitted Westminster Hospital 18/5/56. Treated with digoxin and oxygen	Vomiting. Occasional cyanosis. Poor weight gain	Apex beat in 6th left intercostal space in anterior axillary line. Loud, rasping holosystolic murmur maximal in lower left parasternal area	E.C.D. Grade II. Patent ductus	Died in acute heart failure on 29/5/56
3	9/2/54	Full-term, normal pregnancy. Normal delivery. Birth weight 8 lb. 12 oz. (3,970 g.). Slow weight gain. Pertussis at 7 months. No weight gain after 2 years. Congenital heart disease diagnosed at 2½ years. Admitted Westminster Hospital 7/4/57. Weight 24 lb. (10.9 kg.)	Failure to thrive	Apex beat in 5th left intercostal space in anterior axillary line. Harsh holosystolic murmur over whole praecordium, maximal in lower left parasternal area. Inconstant apical diastolic murmur	E.C.D. Grade II	Cardiac catheterization and selective angiocardiology 9/4/57. Cardiomy on 1/5/57 using Lillehei-deWall type pump oxygenator. Died of irreversible ventricular fibrillation after repair of ostium primum and cleft mitral leaflet
4	28/6/52	Full-term, normal pregnancy. Normal delivery. Birth weight 6 lb. 4 oz. (2,835 g.). Poor weight gain from birth. First admitted Westminster Hospital October, 1952, with pneumonia. Congenital heart disease diagnosed. Numerous subsequent admissions with chest infections and heart failure. Also in hospital elsewhere during infancy with severe gastroenteritis	Failure to thrive. Recurrent chest infections	Aged 4½. Apex beat in 6th left intercostal space in anterior axillary line. Tapping impulse. Parasternal systolic thrill. Loud, rasping holosystolic murmur over whole praecordium, maximal in lower left parasternal area. Apical diastolic murmur. Pigeon chest	E.C.D. Grade I	Died in January, 1958, of heart failure precipitated by pneumonia. Weight at death 23½ lb. (10.7 kg.)
5	17/12/57	Full-term, normal pregnancy. Normal delivery. Birth weight 7 lb. 4 oz. (3,290 g.). Mongol. Rapid breathing from birth with very poor weight gain. First admission to Westminster Hospital 11/8/58	Slow weight gain. Rapid breathing. Mongolism	Puny mongol infant. Apex beat in 5th left intercostal space in mid-clavicular line. Forceful impulse. Soft apical systolic and diastolic murmurs	E.C.D. Grade II	Fully investigated in August, 1958. Readmitted December 15/1/59 under profound hypothermia (Drew's technique). Defects repaired but infant died 24 hours post-operatively of heart-block
6	3/7/52	Full-term, normal pregnancy. Normal delivery. Birth weight 6 lb. 2 oz. (2,780 g.). Poor weight gain from birth. At 17 months admitted to hospital with bronchopneumonia. Congenital heart disease diagnosed. Frequent recurrent chest infections until operation in 1959. In congestive heart failure once at 5	Failure to thrive. Recurrent severe chest infections	Aged 7. Apex beat in 5th left intercostal space in anterior axillary line. Diffuse impulse. Systolic thrill with rasping holosystolic murmur, maximal in lower left parasternal area. Short apical diastolic murmur	E.C.D. Grade I	Fully investigated in 1958. Cardiomy performed 17/9/59 under profound hypothermia (Drew's technique). Ostium primum closed and clefts in mitral and tricuspid valve leaflets repaired. Excellent post-operative progress
7	25/11/56	8 weeks premature. Breech presentation. Caesarean section. Birth weight 4½ lb. (2,040 g.). In incubator for several weeks. Congenital heart disease diagnosed soon after birth. Breathless and failed to thrive until operation in 1959	Failure to thrive. Dyspnoea	Aged 1. Apex beat diffuse. Loud, rasping holosystolic murmur maximal in lower left parasternal area	E.C.D. Grade I	Fully investigated in 1957. Defects successfully repaired in July, 1959, under profound hypothermia (Drew's technique). Now well and putting on weight
8	23/8/51	Full-term, normal pregnancy. Normal delivery. Birth weight 7 lb. 13 oz. (3,345 g.). Normal progress. Heart murmur first heard at routine school examination	None	Aged 7. Apex beat in 5th left intercostal space in anterior axillary line. Blowing holosystolic murmur, maximal in pulmonary and mitral areas. P II widely split. Short early apical diastolic murmur	E.C.D. Grade I	Fully investigated in 1958. Successful repair of defects 8/2/60 under profound hypothermia (Drew's technique)
9	20/9/58	Full-term, normal pregnancy. Forceps delivery. Birth weight 7 lb. 11 oz. (3,485 g.). Mongol. Blue at birth but then well till 4 months, when he weighed 13 lb. 7 oz. (6,095 g.). Subsequently lost weight and was admitted to hospital, where congenital heart disease was diagnosed. Admitted Westminster Hospital 27/4/59	Failure to thrive. Mongolism	Apex beat in 5th left intercostal space in anterior axillary line. Systolic thrill and loud rasping holosystolic murmur maximal in lower left parasternal area. Machinery murmur in Gibson's area	E.C.D. Grade III. Patent ductus	Fully investigated during admission to Westminster Hospital. Died 3 weeks after discharge home in London Hospital of bronchopneumonia and heart failure
10	10/9/59	Full-term, normal pregnancy. Normal delivery. Birth weight 8 lb. (3,630 g.). Labouring breathing from second day. Congenital heart disease diagnosed at 3 weeks and digitalized. Poor weight gain. Admitted Westminster Hospital 24/11/59 with pneumonia and heart failure	Poor weight gain. Labouring breathing. Severe chest infection. Heart failure	Apex beat in 5th left intercostal space in mid-axillary line. Systolic thrill and loud, rasping holosystolic murmur, maximal in lower left parasternal area. Crepitations at bases	E.C.D. Grade II	Investigations interpreted as showing a ventricular septal defect with severe pulmonary hypertension. Died 24 hours after operation of pulmonary arterial banding, 2/1/60
11	26/8/56	Full-term, normal pregnancy. Normal delivery. Birth weight 7 lb. 13 oz. (3,545 g.). From 6 months had recurrent coughs and respiratory illnesses. Easy tiring on walking and often squatted. Breathless on exertion	Easy tiring. Breathlessness. Squatting. Recurrent respiratory illnesses	Aged 2. Apex beat in 5th left intercostal space in anterior axillary line. Harsh systolic murmur, maximal in lower left parasternal area. P II widely split. Apical diastolic murmur	Large ostium secundum	All investigations characteristic of E.C.D. Cardiomy performed 24/9/59 under profound hypothermia (Drew's technique). Large secundum patency closed. Died 10 hours post-operatively of acute right heart failure
12	17/12/56	Illegitimate. No details of birth available. Admitted first to Westminster Hospital 1/2/57. Congenital heart disease diagnosed. Subsequently in Queen Elizabeth Hospital for Children, Banstead, and Westminster Hospital until operation in December, 1959. Progressed reasonably well. Weight at 3 years 28 lb. (12.7 kg.)	Poor feeder in early infancy	Aged 3. Apex beat in 5th left intercostal space just outside mid-clavicular line. Systolic thrill and rasping holosystolic murmur, maximal in lower left parasternal area. Apical diastolic murmur. P II normally split	Large ostium secundum with inferior edge close to fused endocardial cushions	Investigations characteristic of E.C.D. Cardiomy on 17/12/59 under profound hypothermia (Drew's technique). Defect successfully closed. Post-operative condition excellent

other cases and were, in fact, considered radiologically compatible with the mistaken diagnosis of endocardial cushion defect. The x-ray findings are of moderate or considerable enlargement of all heart chambers, with fullness of the pulmonary artery and pulmonary plethora. Where there is significant pulmonary hypertension with a raised pulmonary resistance the characteristic clearing of the periphery of the lung fields is observed (see Figs. 2-4).

FIG. 2.—Chest radiograph of Case 1 with a Grade III endocardial cushion defect showing generalized cardiac enlargement with fullness of the pulmonary artery.

FIG. 3.—Chest radiograph of Case 4 with a grade I endocardial cushion defect and severe pulmonary hypertension. It was taken a few months before death at 5½ years. The patient had suffered from frequent severe respiratory infections. There is biventricular enlargement and gross enlargement of the right atrium and pulmonary artery with pulmonary plethora. The peripheral lung fields are relatively oligæmic.

Electrocardiography

We are in agreement with Toscano-Barbosa *et al.* (1956) in believing that cases of endocardial cushion defect show a highly characteristic electrocardiographic pattern, and have encountered no proved case without it. The features are: (1) in the standard leads—left axis deviation; (2) in the chest leads—right or combined ventricular hypertrophy patterns; (3) in the unipolar limb leads—a mainly negative QRS deflection in aVF, positive in aVL, and a large or small Q wave with a prolonged, often large, R wave in aVR. These features can be seen in the electrocardiograms of all our patients, including those with large low secundum defects (Figs. 5-16).

Reconstruction of the QRS frontal plane vectorial loop shows, in nearly all the cases, a pattern of a mainly anticlockwise loop lying for the most part above the iso-electric point (Fig. 17). This course is the opposite of that usually described in cases of ostium secundum atrial defects.

Certain circumstances may produce modifications in the cardiographic and vectorial patterns. The most important of these is the progressive development of pulmonary hypertension. Fig. 18 shows the changes brought about by mounting pulmonary hypertension in a boy (Case 4) who was observed from early infancy to death in congestive heart failure at 5½ years of age. The left-axis deviation in the standard leads was replaced by right-axis deviation, and the chest leads showed increasing evidence of right ventricular systolic overload. The QRS frontal vector loop inclined further and further to the right.

Cardiac Catheterization Studies

The low course of the catheter in traversing an ostium primum is a useful observation during cardiac catheterization, as pointed out by Wakai *et al.* (1956), but does not exclude an ostium secundum with a low inferior margin.

Haemodynamic data obtained at catheterization in 11 patients (9 endocardial cushion defects and 2 large secundum defects) are shown in Table II. Most, but

FIG. 4.—Chest radiograph of Case 11 with a large ostium secundum patency.

not all, cases showed significant arterialization in the right atrium, and, where this was not demonstrated, arterialization at ventricular level was apparent (Cases 9 and 10). In five cases increments of arterialization were found at atrial and ventricular levels. In all except Cases 7, 10, and 12 there was some degree of systemic arterial desaturation. In Case 7 an arterial oxygen saturation was not determined.

Indicator dilution curves were obtained in several cases at catheterization. We have used a Stanley Cox ear oximeter, Cambridge Instrument Co.'s amplifier, and an Evershed recorder, with methylene blue as the indicator substance in individual injection doses of 15-25 mg., since multiple injections are safe with this rapidly excreted material and we have been interested

only in the configuration of the dye curves. Where possible, curves have been obtained from sequential injections into the two main pulmonary arterial branches. Differentiation of ostium secundum from ostium primum defects by this method has been fully discussed by Wakai *et al.* (1956), who have shown that in most cases of persistency of the ostium secundum there is a preferential left-to-right shunt of right pulmonary venous blood because the orifices of the right pulmonary veins are in close proximity to the septal defect. This gives an indicator dilution curve with a characteristic left-to-right shunt configuration with right pulmonary arterial injection, which is absent or much less marked in the curve obtained with left pulmonary arterial injection. With an ostium primum patency this

TABLE II.—Cardiac Catheterization Data

Case No.	Age	Percentage Oxygen Saturations						Pressures in mm. Hg					Flow in Litres/Minute		Shunts in l./min.		Peripheral Pulm. Resistance Dynes/cm. ²
		Mixed Venous	R.A.	R.V.	P.A.	Systemic Arterial	P.V.	R.A.	L.A.	R.V.	L.V.	P.A.	Sys-temic	Pul-monary	L.-R.	R.-L.	
1	6 weeks	62	90	92	89	94	94	Mean 6	Mean 7	57-5		60/30	1.5	10.0	8.5		319
3	3 years	72	83	84	85	91		" 9		38/0		32/12	3.4	12.5	9.1		141
4	4 "	61	71	77	76	86	89	" 1	" 2.5	62/0		56/25	2.4	5.7	3.6	0.3	575
5	7 months	53	77	85	76	93	95	" 7.5	" 7.5	88/0		83/43	1.0	5.0	4.1	0.1	930
6	5 years	65	77	82	83	91		" 0		50/0		50/5	3.3	10.8	7.5		163
7	13 months	79	85	88	82			" 1.25		72-5		68/35	4.1*	10.0	6.1		353
8	7 years	69	84	89	88	93		" 2	" 2.5	33/0	90/0	32/6	4.0	12.5	8.5		115
9	7 months	58	58	73	76	85		" 0		70-5		74/30	1.9	5.8	4.4†	0.5	687
10	3 "	48	47	66	67	95		" 7		75/0		80/35	1.0	1.7	0.7		2,854
11	2 years	62	80	82	80	93	93	" 12	" 15	63/0	90/0	55/7	2.0	5.7	3.7		450
12	3 "	76	80	87	86	96	97	" 0	" 1	24/0	95/0	20/10	3.0	6.7	3.7		131

* Assuming systemic arterial blood 95% saturated with oxygen. † Assuming pulmonary venous blood 95% saturated with oxygen.

FIGS. 5-16.—Electrocardiograms of the 10 cases of endocardial cushion defect and two cases of ostium secundum patency.

preferential shunting does not occur, since the orifices of all the pulmonary veins are distant from the defect; thus curves from right and left pulmonary arterial injections are similar in form and both show a left-to-right shunt pattern. We have obtained similar curves

in one of our cases of large ostium secundum patency where the lower edge of the defect reached almost to the fused endocardial cushions.

Angiocardiography

Selective angiocardiography was performed on six of the cases of endocardial cushion defect (Cases 1, 3, 4, 7, 9, and 10). Case 9 had two examinations with injection of contrast medium into the left and right ventricles. The investigation was also performed on one of the two cases of large ostium secundum patency (Case 12).

In this total of eight examinations the catheter tip was placed in the right ventricular outflow tract in six instances (Cases 1, 3, 7, 9, 10, and 12), in the right atrium in Case 4, and in the left ventricle (via a ventricular septal defect) in Case 9.

With the exception of Case 7, with a grade I cushion defect, and Case 12, where there was a simple but large ostium secundum patency, all the cases examined with ventricular injection of contrast medium had an inter-ventricular communication and showed simultaneous filling of all heart chambers almost immediately after commencement of the injection. In Case 7, on opacification of the left heart chambers, no opacification of the right atrium or re-opacification of the right ventricle could be detected, although marked re-opacification of the pulmonary arterial tree occurred.

The angiocardiogram in Case 12 demonstrated the atrial communication with passage of contrast medium from the left to the right atrium and subsequent continuing opacification of all the heart chambers throughout the remainder of the study.

In Case 4, where the injection was into the right atrium, there was immediate filling of the other three chambers, which continued through the examination. The right-heart chambers were very large and the lungs showed hypertensive changes with rapid tapering of the pulmonary arterial branches and paucity of opacification in the peripheral lung fields.

A finding in nearly all the cases of endocardial cushion defect to which we attach some diagnostic significance was intense prolonged opacification of an enlarged left atrium. In the lateral films of some of these cases it is possible to see the forceful but incomplete ejection of left atrial contents into the left ventricle during atrial systole and the reverse effect (ejection of left ventricular contents retrogradely through the mitral valve) during left ventricular systole. Owing to the large left-to-right shunt of blood in these cases a constant finding was smallness of the aorta.

Operative and Necropsy Findings

Five cases of endocardial cushion defect (Cases 3, 5, 6, 7, and 8) and both cases of large ostium secundum patency (Cases 11 and 12) were submitted to cardiectomy. After operation three of the former have survived and progressed well with loss of physical signs, as has one of the latter two cases.

Our first case undergoing operation (Case 3) was incorrectly diagnosed before the procedure as a ventricular septal defect with the probable addition of an ostium secundum patency. At operation on May 1, 1957, use was made of an extracorporeal circulation with a bubble oxygenator of Lillehei-DeWall type. The presence of a large right atrium on inspection at thoracotomy, reinforced with further blood-oxygen-saturation estimations, indicated that an interatrial

FIG. 17.—Reconstructed frontal plane QRS vectorial loops.

FIG. 18.—Serial electrocardiograms and QRS frontal vectorial loops of Case 4 showing the sequential changes of increasing pulmonary hypertension.

communication would be found. Right atriotomy was performed and a large ostium primum, with a cleft anterior leaflet of the mitral valve, was repaired. On reversing the potassium-induced heart arrest, however, irreversible ventricular fibrillation ensued. At necropsy, in addition to the defects already stated, there was a high ventricular septal defect (grade II endocardial cushion defect).

The four other cases of endocardial cushion defect and two of ostium secundum patency were operated upon, utilizing the technique devised by Drew (Drew *et al.*, 1959) of profound hypothermia with an extra-corporeal circulation and using the patient's lungs for oxygenation. The correctness of the diagnosis was verified in the cases of endocardial cushion defect, and

satisfactory curative surgical procedures were performed. The first infant operated upon (Case 5) recovered consciousness and was well for two and a half hours but died suddenly of heart-block, as seen on the continuous E.C.G. recording. At necropsy no other satisfactory explanation of death could be found. Good repair of the defects had been achieved and the wounds were healthy and well repaired. The three subsequent operations have all been carried out with rapid post-operative recovery and the absence of any untoward complications. Clinically, the repairs seem to have been curative.

The two cases of ostium secundum patency were incorrectly diagnosed as having endocardial cushion defects. The first patient operated upon (Case 12) died a few hours after closure of the chest wound. The child had a pulmonary resistance of 450 dynes/sec./cm.⁻⁵ and death was attributed to acute right-heart failure. The action of the left ventricle after closure of the atrial defect and re-warming of the patient appeared satisfactory. The other child (Case 12) has made an excellent post-operative recovery and her cardiac signs have disappeared.

On January 1, 1960, we submitted to operation for pulmonary arterial banding a 3-months-old infant (Case 10) who was diagnosed, on catheterization evidence, as having a ventricular septal defect with gross pulmonary hypertension. Congestive heart failure had been treated with digitalis, oxygen therapy, and mercurial diuretics with relief. At thoracotomy the initial pulmonary arterial systolic pressure of 65 mm. Hg was reduced to 35 mm. by the application of a tape round the main pulmonary trunk. After a day of satisfactory recovery from the operation, the infant developed cardiac arrest, which responded to immediate cardiac massage in the ward, but she died an hour later of a second arrest which failed to respond to direct massage of the heart. At necropsy the infant was found to have an endocardial cushion defect of grade II type.

FIGS. 19-21.—Photographs of necropsy specimens from two cases of endocardial cushion defect and one case of large ostium secundum patency. Fig. 19. Case 1, with a grade III endocardial cushion defect. Fig. 20. Case 4, with a grade I defect. Fig. 21. Case 11, with a large atrial defect of ostium secundum type. The defect has been surgically repaired.

Four of our patients with endocardial cushion defect have died in early infancy or childhood without the benefit of surgery. One infant (Case 2) succumbed at 2 months before detailed cardiac investigation could be carried out. She had a grade II defect with the addition of a patent ductus. The other three were all fully investigated and diagnosed before death: the first (Case 1) was 5 months at death and had a grade III defect; the second (Case 4) survived until 5½ years with a grade I defect, aggravated by frequent severe pneumonic illnesses; and the third (Case 9), a mongol, lived for 8 months and died shortly after admission to the London Hospital with pneumonia and congestive failure. We are indebted to Dr. A. D. M. Jackson for kindly allowing us to inspect the heart and confirm our previous diagnosis of endocardial cushion defect (grade III) with a patent ductus.

Photographs of the heart lesions in necropsy specimens of some of these cases are shown in Figs. 19–21. The grade of endocardial cushion defect in the 10 cases is shown in Table I.

Discussion

The rapidly increasing application of intracardiac operative techniques in the corrective treatment of congenital heart defects has resulted in the necessity for precise and accurate pre-operative diagnosis in this field. Surgeons, in order to carry out their procedures, utilize different methods for the open exposure of the heart, depending on the type of lesion they expect to find at operation. This is the case in the operative treatment of ostium secundum atrial defects and endocardial cushion defects. The former can usually be dealt with under moderate hypothermia or through an atrial well, whereas the latter generally need the application of an extracorporeal circulation with or without profound hypothermia.

Considerable attention in many centres, notably in the United States, has been given to this problem of differentiating ostium secundum atrial defects from the more complex endocardial cushion lesions and many diagnostic pointers have been described. There is certainly nothing in symptomatology to separate them apart from the common observation that, as a rule, ostium secundum defects do not produce significant symptoms during infancy and childhood in the majority of cases. In the eight years during which these cases have been observed we have encountered only two deaths in infants or young children due to a simple ostium secundum defect, while four patients with endocardial cushion defect have died (excluding the three post-operative deaths), three of them in the first year of life.

Even the extent of the particular lesion does not seem constantly to determine the severity of the symptoms except that grade III endocardial cushion defects produce early infantile disability and death. Our grade I cases have included a child who was symptomless up to successful operation at 8 years (Case 8) and a child who died at 5 years in congestive heart failure (Case 4). Pulmonary hypertension was present with typical histological changes of hypertension in the lung arterioles. From observations we have made on pulmonary hypertension complicating patent ductus (Anderson and Coles, 1955) and from the effect of pulmonary arterial banding in infants with ventricular septal defects, large pulmonary artery blood flow, and a high pulmonary systolic pressure, we attribute the

hypertensive vascular changes found in the pulmonary arterioles of many cases of left-to-right-shunt defects in association with an elevated peripheral pulmonary resistance to the increased pulmonary blood flow. If we are right in this belief there is hope that in the severely affected infants early surgical correction may either prevent the development of these hypertensive changes in the pulmonary arterioles or lead to a reversal of those which are present. In this context it is highly significant that the lungs of Case 1, who had a grade III cushion defect and died at 5 months in heart failure, did not show hypertensive arteriolar changes.

Children with pulmonary plethora are particularly liable to recurrent chest infections, and these, if severe and frequent, may be extremely crippling and even fatal. Case 4 demonstrates well the adverse effects of an early attack of pneumonia. Although his weight gain from birth was slow, he otherwise appeared normal until 3 months, when he developed pneumonia. His birth weight was 6 lb. 4 oz. (2,835 g.) and weight at 3 months 8 lb. 11 oz. (3,940 g.). Subsequent to this he had repeated pneumonic illnesses, his growth was excessively slow, and at death at 5½ years he weighed 23½ lb. (10.7 kg.).

We have not found physical signs diagnostic in our cases. In many of them the systolic murmur suggested the presence of a ventricular septal defect, especially as it was often maximal in the lower left parasternal area and accompanied by a thrill. The association of an apical diastolic murmur is not infrequent in ventricular defects. This combination of murmurs was heard in five of the cases of endocardial cushion defect and in both cases of large secundum defects where there was no lesion of the mitral valve. It is possible that the diastolic murmur in all these cases results from the passage of blood across the atrial defect or from a torrential flow through the tricuspid valve (Wood, 1956).

Some degree of systemic arterial oxygen desaturation was found in nearly all cases at cardiac catheterization. We regard any saturation below 95% as indicating some degree of desaturation, and this has been confirmed by obtaining arterial saturations in excess of 96% in nearly all those cases we have catheterized in which we have found no shunts between the lesser and greater circulations and no pathological lesions in the lungs. It will be noted that in three of the five instances in which pulmonary venous blood was sampled the saturations were below 95%. A survey of the catheter findings in six cases of simple ostium secundum patency, 21 cases of ventricular septal defect, and nine cases of patent ductus have shown the following results:

Lesion	No. of Cases	No. Showing Arterial Desaturation
A.S.D.	6	5 (83%)
V.S.D.	21	12 (57%)
P.D.A.	9	6 (67%)

It is possible that oxygen desaturation of systemic arterial blood in cases of left-to-right shunt may be due to impaired gaseous interchange across the respiratory epithelium as a result of the increased pulmonary flow, or to the presence of a small but significant right-to-left shunt across the defect, or to the effects of basal narcosis or anaesthetic agents, or to varying combinations of these factors.

There does not appear to be any means of distinguishing certainly between an endocardial cushion

defect and a large low-lying ostium secundum patency, since catheterization data, the course of the catheter in traversing the defect between the atria, and indicator dilution curves obtained from both main pulmonary arterial branches are identical in the two conditions. At the present time, therefore, this type of ostium secundum defect will be diagnosed as an endocardial cushion defect, and operative treatment, if undertaken, will be carried out utilizing some form of extracorporeal circulation. Although this may increase the hazards of operation to individual patients with secundum defects which could have been closed under moderate hypothermia, it is far better that this small minority of cases should undergo an unnecessarily complicated operation than that cases of endocardial cushion defect should be found inoperable owing to the application of inadequate techniques giving the surgeon insufficient time to repair the malformations.

It seems probable that the cases which have been described in the past as ostium primum defects without atrio-ventricular valve involvement are really instances of the type of ostium secundum persistence which we have described in this paper. The deficiency in the interatrial wall is large, relatively posterior, and extends down to within a few millimetres of the fused endocardial cushions, and this latter feature gives it superficially the appearance of an ostium primum defect. The electrocardiograms of our two cases were similar to those found in endocardial cushion defects, and had characteristic frontal vectorial loops. McGoon *et al.* (1959) state that these findings may be observed in 3% of ostium secundum defects and in 13% of ventricular septal defects. We have found the same patterns in one case of patent ductus (Fig. 22). We are inclined to agree

FIG. 22.—Electrocardiogram from a 7-months-old infant with a patent ductus, showing the similarity to the pattern characteristic of endocardial cushion defects.

with Toscano-Barbosa *et al.* (1956) from the Mayo Clinic that the cardiographic pattern probably reflects an abnormal conduction pathway due to the disordered anatomical relationships in the atrio-ventricular region in cases of endocardial cushion defect as well as in the cases of low-reaching ostium secundum patency. It is difficult, however, to see why a similar pattern should result from the usual type of atrial defect and from some ventricular septal defects and patent ductus. An anatomical abnormality of congenital origin cannot be regarded as likely in the atrio-ventricular region in all these cases, and it must therefore be assumed that the electrocardiographic changes are due to hypertrophy of parts of the heart muscle in response to the demands

of the particular defect leading to alterations in the functional capacity of portions of the conduction pathway.

The mortality from endocardial cushion defects is high in infancy and early childhood. For this reason we feel it is desirable to consider early corrective surgery in this group of cases when they present with symptoms and signs of heart disablement. Vigorous medical treatment on orthodox lines should be instituted first and detailed investigation carried out whenever possible to exclude some other lesion mimicking an endocardial cushion defect. Although the risks of intracardiac operation in an infant are great, we believe that, with increased experience in this group of cases, the mortality will diminish to the level in older children.

Summary

An account is presented of the symptoms, physical signs, and haemodynamic data in 10 cases of endocardial cushion defect and two cases of large ostium secundum patency, in one of which the lower edge of the hole nearly reached the fused endocardial cushions. All the cases were seen in infancy or childhood and were fully investigated with the exception of one infant who died at 2 months.

At present there is no certain means of differentiating between an endocardial cushion defect and an ostium secundum of this particular type. Both have similar physical signs, electrocardiographic patterns, and catheterization findings, including a low course of the catheter through the atrial defect, and identical indicator dilution curves when the indicator substance is injected into the right and left pulmonary arteries.

The results of curative surgical operations on the patients so treated are described. Five cases of endocardial cushion defect were operated upon. The first child was incorrectly diagnosed before cardiomy, as a case of ventricular septal defect with a probable additional ostium secundum patency. Open heart surgery was effected in this case in May, 1957, using a bubble oxygenator of Lillehei-DeWall type, and the child did not survive the procedure. The four other cases were correctly diagnosed and submitted to cardiomy, using the technique of Drew—namely, a combination of extracorporeal circulation and profound hypothermia. Three of these patients have survived and are clinically cured.

The two cases of ostium secundum patency were erroneously thought to have endocardial cushion defects, but at operation, using profound hypothermia, the true nature of the lesion was recognized and the defect appropriately repaired. The first patient operated on died from acute heart failure a few hours after closure of the chest; the second has done well and appears to be clinically cured.

Final diagnosis in all 12 cases has been made on visual inspection at operation or necropsy.

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MANAGEMENT OF CASES OF ABORTION

BY

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This review is based on the work of the abortion unit at Walton Hospital, Liverpool, during the years 1958 and 1959, and the results achieved with a particular type of treatment in cases of abortion admitted to it.

Cases of abortion are usually admitted to a gynaecological ward, a system which is very inconvenient when large numbers are admitted annually. Among its disadvantages is the blocking of a variable number of beds, making the arrangements of clinic admissions difficult, and there is also the frequent use of an operating theatre at inconvenient times.

In January, 1950, at this hospital, a separate ward block with 40 beds was made available as an abortion unit. It is arranged in three floors, with an operating theatre on the first floor. Patients admitted with a diagnosis of abortion are taken to the abortion unit unless they require resuscitation in the admission room.

Criteria Used in Classification

An abortion was regarded as threatened if the cervix was closed and the bleeding slight. In inevitable abortion the cervix must be open with the products of conception protruding and felt digitally or seen by direct vision. No abortion was regarded as complete unless the intact ovum or the foetus followed by an intact placenta was seen by the hospital medical officer.

Difficulty often arose in differentiating between threatened, incomplete, and complete abortion. The amount of bleeding was not a reliable guide; in some cases the pregnancy continued after relatively heavy bleeding, while in others a slight loss of blood may have been followed by the unrecognized expulsion of an intact ovum. When comparison of the period of amenorrhoea with the size of the uterus, the state of the cervix, and examination of the vaginal discharge for fragments of

ovum still left the diagnosis in doubt, the abortion was regarded as threatened. Biological pregnancy tests and a period of observation helped to establish the diagnosis later.

The criteria for the diagnosis of septic abortion were similar to those adopted by Stallworthy (1948)—namely, that an abortion was regarded as septic when associated with pyrexia for which no other cause was found, with offensive purulent discharge, or with evidence of pelvic infection.

Treatment

Cases are admitted to the ground-floor wards and a vaginal examination is made. If the abortion is inevitable the part of the ovum in the cervical canal is removed either digitally or, if this is impossible, with a pair of sponge forceps under direct vision. Ergometrine 1 mg. is given intramuscularly. Cases of incomplete abortion are also given ergometrine if there is any bleeding.

Each week, operating sessions are held on Tuesday, Thursday, and Saturday mornings. The day before operation all cases in which surgical evacuation of the uterus is to be performed are taken to the first-floor ward. They include all the incomplete abortions and any "complete" abortions where vaginal bleeding or discharge persists.

Morphine sulphate 1/6 gr. (11 mg.) and atropine sulphate 1/100 gr. (0.65 mg.) are given intramuscularly as premedication one hour before operation. Thio-pentone alone is used in most cases, nitrous oxide and oxygen being used occasionally for supplementary maintenance of anaesthesia. Vulval toilet is carried out, using 1% cetrimide solution. Retained products of conception are removed digitally or by sponge forceps, and curettage is performed with a large, sharp curette. If bleeding is troublesome ergometrine 1 mg. is given intravenously. Packs are not left in the uterus and intrauterine douches are not used.

The patient is moved to the second floor on the day after operation, and if her progress is satisfactory she is discharged on the fourth day after operation.

This scheme of treatment has been in operation for nine years and the results have been very satisfactory.

The Investigation

It is estimated that about 15 to 20% of pregnancies terminate in abortion, and during the two-year period, January, 1958, to December, 1959, 2,387 patients were admitted to the abortion unit (Table I). The district from which these patients are drawn is a large over-crowded area adjacent to the docks of Liverpool. The general standard of health and hygiene is low and anaemia is common.

TABLE I.—Admissions During 1958 and 1959

Total no. of admissions	2,387
No. booked (at hospital antenatal clinic)	204
No. not booked	2,183
Average age	28.9 years
Primigravidae	24.8%
Unmarried	5.1%
Average maturity at onset of abortion	11.7 weeks
Less than 16 weeks' gestation	87%
Prophylactic rest (habitual abortions)	56
Hydatidiform moles	3
Ectopic gestation	1

The average age of the patients was 28.9 years, the youngest being 15 and the oldest 54. This corresponds with other series in the literature; in 2,665 cases quoted by Davis (1950) it was 29 years. As he points out,