Congenital heart disease among 160 480 liveborn children in Liverpool 1960 to 1969

Implications for surgical treatment

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SUMMARY Among 160 480 children born alive between 1960 and 1969 in Liverpool, 884 patients with structural congenital heart disease were identified. Data on these patients have been reviewed in order to estimate the number likely to need cardiac surgery during childhood and adolescence. Though only 33-9% of patients had surgery, we estimate that if current policies for management were followed, 475 (53-7%) patients would now require surgery.

Extrapolation of this data suggests that each year in England and Wales approximately 830 infants (1383 per million livebirths) will require cardiac surgery within the first year of life and a further 1424 operations (2374 per million livebirths) will be required in later childhood or adolescence. No attempt has been made to estimate the number of operations for congenital heart disease which may prove necessary in adult life.

Although the incidence of congenital heart disease has been documented by many authors, these studies have concentrated on the epidemiological aspects of the problem, and little attempt has been made to assess the proportion of cases in an unselected population of patients with congenital heart disease who might require surgical treatment. Previous estimates of the resources needed for the optimum care of children with congenital heart disease are therefore likely to be based on data from selected clinic populations which may not necessarily reflect the true situation. An unselected group of patients with congenital heart disease born between 1960 and 1969 in the Liverpool area and originally reviewed by Kenna et al. have been studied in order to determine the proportion of cases who have undergone operation or who might have benefited from operation when account is taken of the advances in management and improved prospects for survival which have taken place in the past decade.

Subjects and methods

The Liverpool Registry of Congenital Malformations contains records of all children with congenital malformations who were born to residents of the Registry area (the cities of Liverpool and Bootle). The methodology of the Registry has been described in detail previously. One thousand one hundred and nineteen patients out of a total of 160 480 children born during the decade 1960 to 1969 were registered as having a congenital cardiac anomaly. The hospital records of these patients have been reviewed with a view to determining whether surgery was actually carried out or would have been considered necessary by present criteria in those patients who died without surgical intervention. Where necessary, the diagnosis of individual patients has been revised. Eight hundred and eighty four liveborn patients were identified with definite structural congenital heart disease, though a final diagnosis had not been reached in 29 cases. Table 1 shows the number of patients and the incidence per 10 000 livebirths for each of the major diagnostic categories.

Fifteen patients with non-structural cardiac defects (including cases of endocardial fibroelastosis, congenital heart block and non-obstructive cardiomyopathy) and eight patients with situs inversus and dextrocardia and a normal heart were not included. Thirty-five stillborn infants with congenital heart disease, and 177 cases in whom subsequent examination afforded insufficient evidence of congenital heart disease were also excluded. Most of these patients are now considered to have innocent systolic murmurs.

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Table 1  Number of patients and incidence per 10 000 livebirths in each major diagnostic category compared with incidence of these lesions in three other recent studies

<table>
<thead>
<tr>
<th>Lesion</th>
<th>No. of cases</th>
<th>Incidence per 10 000 live births</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Liverpool</td>
<td>Hoffman</td>
</tr>
<tr>
<td>VSD</td>
<td>287</td>
<td>18-0</td>
</tr>
<tr>
<td>PDA</td>
<td>105</td>
<td>6-5</td>
</tr>
<tr>
<td>ASD</td>
<td>52</td>
<td>3-2</td>
</tr>
<tr>
<td>AVD</td>
<td>21</td>
<td>1-3</td>
</tr>
<tr>
<td>PS</td>
<td>67</td>
<td>4-2</td>
</tr>
<tr>
<td>AS</td>
<td>45</td>
<td>2-8</td>
</tr>
<tr>
<td>Coarctation</td>
<td>56</td>
<td>3-5</td>
</tr>
<tr>
<td>TGV</td>
<td>44</td>
<td>2-7</td>
</tr>
<tr>
<td>Fallot</td>
<td>52</td>
<td>3-2</td>
</tr>
<tr>
<td>Pulm atresia</td>
<td>7</td>
<td>0-4</td>
</tr>
<tr>
<td>Tric atresia</td>
<td>15</td>
<td>0-9</td>
</tr>
<tr>
<td>TAPVC</td>
<td>12</td>
<td>0-7</td>
</tr>
<tr>
<td>UVH</td>
<td>15</td>
<td>0-9</td>
</tr>
<tr>
<td>Truncus</td>
<td>10</td>
<td>0-6</td>
</tr>
<tr>
<td>HLHS</td>
<td>25</td>
<td>1-6</td>
</tr>
<tr>
<td>Misc</td>
<td>42</td>
<td>2-6</td>
</tr>
<tr>
<td>Undiagnosed</td>
<td>29</td>
<td>1-8</td>
</tr>
<tr>
<td>Total</td>
<td>884</td>
<td>55-1</td>
</tr>
</tbody>
</table>

Abbreviations: VSD, ventricular septal defect; PDA, persistent ductus arteriosus; ASD, atrial septal defect (secundum); AVD, atroventricular defect (including ostium primum defect); PS, pulmonary stenosis; AS, aortic stenosis; TGV, transposition of the great vessels; Pulm atresia, pulmonary atresia (with intact ventricular septum); Tric atresia, tricuspid atresia; TAPVC, total anomalous pulmonary venous connection; UVH, univentricular heart; Truncus, truncus arteriosus; Misc, miscellaneous; HLHS, hypoplastic left heart syndrome.

*These figures include a variety of "non structural" congenital abnormalities not included in the Liverpool total (for example, congenital heart block, endocardial fibroelastosis, cardiomyopathy, "dextrocardia").

The method of diagnosis by three mutually exclusive categories for each of the major diagnostic groups is shown in Table 2.

In assessing the number of patients who would now be regarded as potential candidates for operation, we have included all those patients who have already undergone operation except in the case of eight patients with a ventricular septal defect who are discussed separately. Those patients dying as a result of their cardiac defect are also included, except when the presence of major non-cardiac malformations would have been likely to preclude a successful outcome. Conversely, three patients who died from unrelated causes are regarded as potential surgical candidates as their survival would have been probable by present-day standards of management. An attempt has been made to group surgical candidates into two age groups—those requiring surgery under 1 year and those requiring operation later in childhood. This grouping has been based on the age of the patient at operation or death, taking into account recent trends towards earlier elective operation for some defects.

Results

Eight hundred and eighty four patients among 160 480 livebirths (55·1 per 10 000) were considered to have definite evidence of congenital heart disease. Table 3 summarises the data relating to the follow up of these patients, and provides an estimate of the number of patients in each diagnostic category who would be considered as surgical candidates according to our present criteria. The age at presentation of the patients is shown from Table 4.

The total number of patients who have undergone operation (either palliative or corrective) in this series up to the present time is 300 (33·9%). Two hundred and ninety-seven patients (33·6%) have died. In 65 cases (7·4%), death followed operation, while in 55 cases (6·2%), death was unrelated to the presence of the cardiac defect. The cause of death in these cases is shown in Table 5. A further 289 patients (32·7%) are alive having had no surgical treatment. The majority of these patients have minor defects, but five patients with Down's syndrome and complete atroventricular canal defects and 12 patients in the miscellaneous category have been included among those with defects which will almost certainly require an operation during adolescence or early adult life. A further six patients have the Eisenmenger syndrome (two with persistent ductus, two with transposition, and one each in the categories of univentricular heart and truncus arteriosus). It is probable that some of these patients, particularly those with persistent ductus, could have been treated surgically had they been recognised earlier.

GENERAL IMPLICATIONS FOR SURGICAL TREATMENT

Of the patients presenting with evidence of congenital heart disease during the first year of life, we expect only a small number of patients dying in the immediate...
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Table 3 Clinical course of 884 liveborn patients with structural congenital heart disease together with an estimate of numbers who might have benefited from surgical treatment by present standards

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Liveborn</th>
<th>Cardiac death no operation</th>
<th>Unrelated death</th>
<th>Underwent operation*</th>
<th>Lost</th>
<th>Alike no operation</th>
<th>Estimated No of potential candidates</th>
<th>Age at first operation under 1 year</th>
<th>Total No. of operations</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD 287</td>
<td>15</td>
<td>24</td>
<td>37 (9)</td>
<td>26</td>
<td>185</td>
<td>(90)</td>
<td>44</td>
<td>17</td>
<td>27</td>
</tr>
<tr>
<td>PDA 105</td>
<td>7</td>
<td>2</td>
<td>85 (3)</td>
<td>0</td>
<td>6</td>
<td>94</td>
<td>20</td>
<td>74</td>
<td>95</td>
</tr>
<tr>
<td>ASD 52</td>
<td>2</td>
<td>6</td>
<td>37 (1)</td>
<td>3</td>
<td>4</td>
<td>41</td>
<td>2</td>
<td>39</td>
<td>41</td>
</tr>
<tr>
<td>AVD 21</td>
<td>9</td>
<td>0</td>
<td>7 (3)</td>
<td>0</td>
<td>5</td>
<td>21</td>
<td>15</td>
<td>6</td>
<td>25</td>
</tr>
<tr>
<td>Coarctation 56</td>
<td>18</td>
<td>4</td>
<td>32 (5)</td>
<td>2</td>
<td>0</td>
<td>53</td>
<td>30</td>
<td>23</td>
<td>65</td>
</tr>
<tr>
<td>PS 67</td>
<td>2</td>
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<td>13 (0)</td>
<td>3</td>
<td>23</td>
<td>19</td>
<td>4</td>
<td>15</td>
<td>21</td>
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<td>Fallot 52</td>
<td>12</td>
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<td>35 (17)</td>
<td>0</td>
<td>0</td>
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<td>20</td>
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<td>65</td>
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<td>TGV 44</td>
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<td>15 (13)</td>
<td>0</td>
<td>3</td>
<td>43</td>
<td>40</td>
<td>3</td>
<td>60</td>
</tr>
<tr>
<td>Tricuspid atresia 15</td>
<td>11</td>
<td>1</td>
<td>3 (2)</td>
<td>0</td>
<td>0</td>
<td>14</td>
<td>12</td>
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<td>25</td>
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<td>Pulmonary atresia 7</td>
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<td>12</td>
<td>12</td>
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<td>12</td>
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<tr>
<td>UVH 15</td>
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<td>13</td>
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<tr>
<td>Truncus 10</td>
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<td>0</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Miscellaneous 42</td>
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<td>4</td>
<td>11 (6)</td>
<td>0</td>
<td>12</td>
<td>34</td>
<td>19</td>
<td>15</td>
<td>43</td>
</tr>
<tr>
<td>Undiagnosed 29</td>
<td>2</td>
<td>0</td>
<td>26</td>
<td>1</td>
<td>4</td>
<td>4</td>
<td>2</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Total 884</td>
<td>177</td>
<td>55</td>
<td>300 (65)</td>
<td>63</td>
<td>289</td>
<td>475</td>
<td>222</td>
<td>253</td>
<td>573</td>
</tr>
</tbody>
</table>

*Includes both palliative and corrective operations. The figure in parentheses indicates the number of patients who died after operation.

†Figure in parentheses indicates number of patients in whom the ventricular septal defect has closed spontaneously.

**Note:** The figure for the total number of operations required in each diagnostic category has been reached by estimating the number of patients within each category who might require more than one operation during childhood. All those patients in the series who actually had a second operation are included and an attempt has been made to estimate the number of second operations needed by those patients with complex defects who died without surgery but who would now be considered eligible for surgical treatment.

Abbreviations as for Table 1.

The postnatal period of obvious major non-cardiac malformations or birth trauma would not require assessment at a paediatric cardiac centre. Twenty-four patients fell into this category and, therefore, 643 of the 667 patients presenting within the first year of life would probably now be referred for assessment. On the basis of the provisional diagnosis at the time of referral, together with the presence or absence of symptoms, it is estimated that 408 (2.5 per 1000 livebirths) of these patients would now undergo full investigation including cardiac catheterisation. Two hundred and twenty-two (54.4%) of these would be potential candidates for operation within the first year of life. Forty-nine patients (5.5%) presenting during the first year of life with major non-cardiac malformations or perinatal problems and 25 patients (2.8%) with hypoplastic left heart syndrome were not included as valid candidates for surgical treatment.

Although only 33.9% of the patients in this series actually underwent operation, it is estimated that a total of 475 (53.7%) would benefit from surgical treatment by present-day standards of management. The results in the individual diagnostic categories and the implications for surgical treatment in each group are discussed below.

**VENTRICULAR SEPTAL DEFECT**

Thirty-seven patients in this group underwent surgical treatment, but in eight cases the preoperative cardiac catheterisation data showed a normal pulmonary artery pressure and a pulmonary to systemic flow ratio of less than 2:0. Only 29 of these patients are therefore regarded as requiring surgery by our present criteria. It is recognised that for units having a more active surgical policy for children with normal or near-normal pul-
monary pressure and moderate sized left-to-right shunts this figure may underestimate actual numbers. A further 15 patients dying with symptoms related to the ventricular septal defect are included as potential surgical candidates, though it should be noted that seven of these cases had Down's syndrome. Limited information was available on the 26 patients lost to follow up but it is unlikely that any of these cases would have required surgical treatment. Spontaneous closure of the ventricular septal defect was noted in 90 of the 185 patients remaining alive without surgical treatment. The remaining 95 patients, at the time of their last outpatient visit, had signs of a small ventricular septal defect and are not considered as potentially surgical candidates.

**ATRIAL SEPTAL DEFECT**

Forty-one patients are regarded as potential surgical candidates. This figure includes 37 patients who underwent operation, two who died without an operation and two lost to follow-up in whom the available data suggest that an operation would have been needed. Operation was not recommended in four cases; in three cardiac catheterisation showed only small defects with pulmonary to systemic flow ratios of less than 1:5 to 1, and in one patient severe mental retardation was present. It is accepted that views may differ on the advisability of operation for children with very small atrial shunts and that a small increase in the number of prospective surgical candidates might be considered appropriate by some units.

**PERSISTENT DUCTUS ARTERIOSUS**

Ninety-four patients in this group were considered to be surgical candidates, including the 85 who underwent operation, seven who died in infancy without surgical treatment, and two living children with the Eisenmenger syndrome. Spontaneous closure of an isolated persistent ductus was recorded in only four patients.

**ATRIOVENTRICULAR DEFECT**

Seventeen of the 21 patients in this group had Down's syndrome. All these patients had complete atrioventricular canal defects. The four normal children had ostium primum defects and underwent successful surgery. All these 21 patients, including those with Down’s syndrome, are included as potential surgical candidates. Since nine patients with complete atrioventricular canal defects died before the age of 15 months, it is considered that most would require an initial operation during the first year of life. Only the four patients with ostium primum defects and two with complete atrioventricular canal defects and moderate infundibular stenosis would be expected to have their first operation after the age of 1 year.

**COARCTATION OF AORTA**

The total number of 56 patients includes 36 with an isolated coarctation and 20 with an additional defect. The three patients not included as potential surgical candidates all died in the early neonatal period, two with a major central nervous system malformation and the third after a tentorial tear during delivery. A fourth patient who died at the age of 9 months during the course of an operation for rectal atresia is regarded as a candidate for cardiac surgery. Two patients who moved from the area early in life had definite signs of coarctation and though we have been unable to locate them, seem probable surgical candidates. Six children (19% of those undergoing operation) required a second operation for recurrent coarctation. The use of an angioplastic repair using the left subclavian artery would probably reduce this recurrence rate substantially, but the improved survival among those patients with additional intracardiac lesions would probably result in a number of second operations using cardiopulmonary bypass to correct these defects. We estimate the total number of operations required as 65.

**PULMONARY STENOSIS AND AORTIC STENOSIS**

In these groups the number of potential surgical candidates includes only those patients actually operated on plus a small number dying in infancy without operation in whom severe stenosis was found at necropsy. Two patients in each group underwent a second operation in childhood and these operations are included in the total number of operations required. No attempt has been made to estimate the number of operations which might be necessary in adult life, particularly in the case of aortic stenosis.

**MAJOR CYANOTIC LESIONS**

In the diagnostic categories of Fallot’s tetralogy, transposition, tricuspid atresia, pulmonary atresia, with intact ventricular septum, total anomalous pulmonary venous drainage, univentricular heart, and truncus arteriosus, all patients have been assumed to require operation unless multiple major non-cardiac malformations were present. Only five cases have been excluded on this basis. In the case of Fallot’s tetralogy, two of the five unrelated deaths were probably avoidable by present standards of management and both are regarded as potential surgical candidates. The number of patients in each group needing surgical intervention within the first year of life reflect the number of patients who underwent operation within this age range, plus the number dying without surgical treatment before their first birthday. It is possible, however, that the estimated total number of operations required might vary considerably from that suggested in Table 3 since this figure will clearly be dependent on...
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the success of the early management of these complex lesions.

MISCELLANEOUS

Included among the 42 patients in this group are six patients with "classical corrected transposition", six patients with complex defects associated with situs ambiguous, seven patients with mitral valve anomalies, four patients with Ebstein's anomaly, three patients with vascular rings, and 16 patients mainly with complex defects difficult to categorise except on an individual basis. Thirty-four patients have been regarded as potentially needing surgical treatment before the age of 16.

UNDIAGNOSED

Twenty-nine patients had clear evidence of a structural cardiac defect but no definite diagnosis had been reached. Twenty-six of these patients had been lost to follow up and in the majority of these cases the provisional diagnosis at the time of their last clinic visit was either a small ventricular septal defect (six cases) mild pulmonary stenosis (11 cases), or mild aortic stenosis (five cases). It is impossible to make an accurate assessment of the number of patients who might have needed surgical treatment if follow up data had been complete. Two patients dying within the first year of life, however, in whom necropsy data were imprecise are regarded as potential surgical candidates as are two patients who have moved from the Liverpool area with a provisional diagnosis of Fallot's tetralogy in one case and ostium primum defect in the other and in whom up-to-date information has not been obtained.

Discussion

A study of congenital heart disease which is based on data collected by a central registry may underestimate the true incidence of some lesions, since symptom-free patients may not come to medical attention. A comparison of the incidence of the major diagnostic groups in this series with that of more intensive prospective studies suggests some patients were indeed lost (Table 1). In the case of ventricular septal defect and pulmonary stenosis, though this shortfall is important from an epidemiological viewpoint, we believe, for the reasons outlined below, that it is unlikely that a significant number of cases who would have benefited from surgical treatment have been lost. The methodology of the registry provides for the regular scrutiny of necropsy records of the children's hospitals and maternity units within the Registry area. The necropsy rate in these units during the period under review was high, and it is therefore unlikely that patients dying with congenital heart disease in infancy would have been missed. Patients with obvious signs or symptoms of congenital heart disease would be referred to one of the two children's hospitals in the area, and the only patients with ventricular septal defect who might have escaped detection in significant numbers are those with small defects which closed early in infancy and those presenting later in childhood when the family were no longer resident within the Registry area. Bearing in mind the high incidence of spontaneous closure of ventricular septal defects in the early years of life and the fact that asymptomatic patients presenting later in childhood almost invariably have small defects, it seems probable that any patients with ventricular septal defect missed are almost certain to be those who would not require surgery.

In the case of pulmonary stenosis, published studies show a considerable variation in incidence (Table 1). When strict diagnostic criteria are applied to the patients undergoing cardiac catheterisation (gradient >25 mmHg) the incidence of this lesion is low, and it seems probable that the cases missed by the Registry fall mainly in the group with small pressure gradients who would not be regarded as requiring surgical treatment in childhood.

With atrial septal defect and coarctation of the aorta the shortfall of cases is more significant. Children with these lesions, because of the absence of symptoms and often unimpressive physical signs, may present later in childhood, and the identification of these two lesions has undoubtedly been affected adversely by the movement of population out of the Registry area during the 1960's and early 1970's. In the case of atrial septal defect, however, the implications for the surgical treatment may not be as great as the degree of shortfall at first suggests. In the study of Mitchell et al seven of the 31 liveborn patients with an atrial septal defect died during the neonatal period. A similarly high proportion of cases was diagnosed at necropsy in the study of Bound and Logan. Since symptoms are uncommon in an uncomplicated atrial septal defect in infancy it seems probable that these patients died of unrelated causes rather than as a result of the cardiac lesion, and they would not therefore be regarded as surgical candidates. Indeed the strikingly high incidence of atrial septal defect in stillbirths and neonatal deaths in some series suggests some overdiagnosis of this defect, possibly related to the widely patent foramen ovale often seen at this age. Nevertheless it must be accepted that our figure probably underestimated the number of patients with an atrial septal defect requiring surgical treatment by perhaps 15 to 20 cases. An adjustment of the figure for coarctation of the aorta of a similar order is also probably necessary. It seems probable that almost all of these cases in both diagnostic groups would have required their initial operation above the age of 1 year, as patients symptomatic in infancy are unlikely to have eluded registration.
RECENT ADVANCES
Recent surgical advances\textsuperscript{12–15} have considerably improved the prognosis for a number of complex congenital cardiac lesions, particularly in infancy. In assessing the number of patients who might benefit from surgical treatment today, we have followed our own current practice in the management of these conditions. An aggressive approach to the management of lesions such as single ventricle, tricuspid atresia, truncus arteriosus, and complex transpositions has been assumed, and major malformations outside the cardiovascular system have not necessarily been regarded as a contraindication to treatment if a favourable outcome seemed possible. Patients with Down’s syndrome have not been excluded from consideration for surgical treatment. It is only in the categories of ventricular septal defect and atrioventricular defect, however, that the number of patients with Down’s syndrome is sufficiently large to make an appreciable difference to the total number of patients requiring surgical treatment. Within these two categories it should be noted that 26 children with Down’s syndrome have been regarded as potential surgical candidates, the majority needing operation within the first year of life.

A recent study from Toronto\textsuperscript{7} has documented a considerable increase in the number of infants with a persistent ductus arteriosus referred during 1975 as compared with 1965. This increase is probably related to the improved prospects for survival in the very premature infant. A similar increase might be expected in this country, but would not necessarily result in an increased number of operations for persistent ductus arteriosus during the first year of life since pharmacological treatment may also be effective in closing the ductus.\textsuperscript{16} Failure of the ductus arteriosus to respond to indomethacin therapy, however, has been documented by a number of authors\textsuperscript{17,18} and some increase in the number of infants requiring surgery for this condition therefore seems inevitable. At present it is impossible for us to assess the possible size of this increase.

The total number of operations needed by the estimated 475 potential surgical candidates is difficult to predict. Our figure of 573 (Table 3) includes all those patients in the series who actually had a second operation. In addition, it includes an estimated figure for the number of second operations likely to be necessary in patients who died without surgery but who would now be considered eligible for an initial palliative operation followed by a definitive procedure at a later date. Only one subsequent operation has been allowed for each patient. Though this may be an underestimate of the requirements for some patients with complex defects, this will probably be offset by the mortality associated with the initial procedure in others. We have not attempted to estimate the number of operations which might become necessary during adult life. It seems probable that the total number of operations needed during childhood and adolescence by the 475 potential surgical candidates would be approximately 573, of which 222 would be within the first year of life. About 40 to 50% of the operations in infancy would be open-heart procedures.

These figures may be used to provide an estimate of the number of operations on children and adolescents with congenital heart disease needed annually in England and Wales. In recent years the total number of liveborn infants in England and Wales has varied from 569 259 in 1977 to 638 028 in 1979. Using 600 000 live births as a basis for calculation and adding 15 patients to our figures in the categories of both atrial septal defect and coarctation of the aorta, to allow for under-estimates of these defects, it appears that each year in England and Wales 1888 of the children born should require an operation for congenital heart disease. The total number of operations needed each year should be 2254, 830 of them needed in infants less than 1 year old. The estimated number of operations required in each diagnostic category (expressed as a number per million livebirths) is shown in Table 6.

\begin{table}
\centering
\caption{Estimated number of operations for congenital heart disease required annually in England and Wales}
\begin{tabular}{|l|l|l|}
\hline
\textbf{Lesion} & \textbf{Estimated annual no. of operations per million livebirths} & \\
& \textbf{Under 1 year of age} & \textbf{Over 1 year of age} \\
\hline
VSD & 106 & 194 \\
PDA & 125 & 467 \\
ASD & 12 & 536 \\
AVD & 93 & 62 \\
Coarctation & 187 & 312 \\
PS & 19 & 100 \\
AS & 25 & 106 \\
Fallot & 125 & 280 \\
TGV & 249 & 125 \\
Pulm atresia & 44 & 50 \\
Tic amereus & 75 & 81 \\
TAPVC & 75 & 0 \\
UVH & 62 & 62 \\
Truncus & 56 & 37 \\
Undiagnosed* & 12 & 12 \\
Misc & 118 & 150 \\
\hline
Total & 1383 & 2374 \\
\hline
\end{tabular}
\end{table}

*See text. Abbreviations as for Table 1.

It is of interest that the estimated figure for the total number of operations needed, at 3759 per million livebirths, is very close to the figure of 3649 per million livebirths calculated from the recommendations of the inter society commission for heart disease resources\textsuperscript{4} for the USA. Our data suggest, however, that relatively fewer operations (1383 per million livebirths) would be required during the first year of life than the 1988 per
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millions of livebirths suggested by Engle et al.4 Our figure is considerably closer to the figure of 850 infant operations (1409 per million livebirths) calculated by Macartney19 from the data collected by the New England regional infant cardiac programme.20 It is also of interest that our figure for the number of patients requiring cardiac catheterisation in infancy, at 2.5 per 1000 livebirths, is very similar to the figure of 2.7 per 1000 livebirths quoted by Macartney.19

In view of the relatively low incidence (5-4 cases per 1000 livebirths) of congenital heart disease in the population studied, caution should be exercised in extending our results to predict the number of patients in England and Wales who might benefit from surgical treatment each year. The surgical requirements of the cases missed by the registry study are unlikely to be the same as those for the patients identified. For the reasons discussed previously, however, it seems probable that, except for atrial septal defect and coartation of the aorta, the majority of the patients missed would not have required operation. Infants presenting and needing investigation and treatment within the first year of life, are unlikely to have been overlooked. An annual figure of close to 850 operations within the first year of life, therefore, seems appropriate for the needs of the present population of England and Wales, assuming the birth rate to remain at its present level. An aggressive approach to the palliation of complex defects together with an increased probability of survival for these cases may result in an increased number of patients requiring second, or even third and fourth operations, later in childhood. Furthermore, our figures make no provision for the number of operations which might be needed for the insertion and replacement, when necessary, of permanent cardiac pacemakers. For these reasons our estimate of 1424 operations needed annually, in patients over 1 year of age, should probably be regarded as a minimum requirement.

Actual estimates of the number of patients undergoing operation for congenital heart disease in England and Wales suggest that in excess of 3000 operations were carried out in 1977 (T English, personal communication). The discrepancy between this figure and our estimate of requirements is probably related to four factors. Firstly, patients in the older age groups in 1977 would have been born at a time when the birth rate was much higher. Secondly, changes in surgical techniques and policy have meant that some older children, adolescents, and even young adults may have been operated on, who, by present criteria, would have had corrective surgery electively earlier in childhood. Thirdly, an increased surgical throughput in many units may have been a transient phenomenon associated with a backlog of patients on the waiting list. Fourthly, patients from outside the UK are included in the actual figures for 1977 and in some centres may have constituted a significant proportion of the total caseload.

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References


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