Structural heart disease in the newborn

Changing profile: comparison of 1975 with 1965

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SUMMARY The earlier detection and investigation of babies with congenital structural heart disease has resulted in earlier treatment and better management of these patients. In 1965, at the Hospital for Sick Children, Toronto, 121 cases were assessed and treated; by 1975 this figures had risen to 226 cases. Few changes were noted in the incidence of the 10 most common malformations, except for the obvious increase in the incidence of patent ductus arteriosus due to survival of increasingly premature infants. The sick cardiac neonate was referred earlier in 1975, studied with newer non-invasive investigations, and, according to the severity of symptoms and signs, was sent sooner to cardiac catheterisation. 80% of babies presenting with cyanosis survived the first month and 60% of babies with congestive heart failure survived. With better surgery and perioperative care, the survival rate in the first month after surgery rose from 37% in 1965 to 70% in 1975.

During the last decade there has been a wider recognition of the need for early referral and aggressive management of the newborn with cardiac malformation (Fyler, 1968; Varghese *et al.*, 1969). The increasing complexity of diagnostic procedures and support services, and the use of specialised surgical techniques (Mustard *et al.*, 1970; Edmunds *et al.*, 1972; Barratt-Boyes *et al.*, 1973), have resulted in a trend to regionalise these efforts (Fyler *et al.*, 1972; Nadas *et al.*, 1973; D. Cook *et al.*, 1978, unpublished data). Consequently, the numbers of patients admitted for this type of care have increased. This hospital reported that 211 neonates with congenital heart disease were assessed retrospectively for the period 1953-57 (Rowe and Cleary, 1960). By 1965, 121 neonates with heart malformation were being seen yearly.

In this paper the presenting features, type of

anomaly, investigative techniques, and results of treatment in newborn patients with heart malformations seen in 1965 are compared with a prospective group of 226 such babies seen in 1975.

Materials and methods

Newborn infants seen with congenital heart malformations at this hospital in 1965 were analysed retrospectively by searching our data bank for infants with cardiac malformations; this data bank has been in operation since 1952 (Rose *et al.*, 1972).

Patients seen in 1975 were evaluated prospectively as neonates requiring cardiological consultation at the hospital. The newborn was first seen by a fellow in cardiology and shortly afterwards by a staff cardiologist. Preliminary investigations, apart from a physical examination, included blood-gas measurements, a chest x-ray, and an electrocardiogram. Echocardiograms were obtained in 128 of the 226 patients with structural heart disease. Results of the preliminary investigations and the recommended treatments were communicated to the appropriate unit staff. Further investigation by cardiac catheterisation and angiocardiology was performed after such consultations in 117 patients. Subsequent evaluations on the ward were performed by cardiologists usually daily, but more or less often according

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to need. Follow-up after discharge, arranged through the referring physician, depended on the severity of the residual cardiac anomaly and the likelihood of future problems.

The final diagnosis was established at necropsy, surgery, cardiac catheterisation, or by clinical assessment only, in descending order of certainty. If multiple defects existed, the most important malformation was listed. A few broad categories of diagnosis were used. For example, hypoplastic left heart syndrome included aortic valve atresia, mitral valve atresia, and hypoplasia of all left heart structures without atresia. In the group hypoplastic right heart syndromes were lesions with hypoplasia of the right ventricle, particularly pulmonary atresia with intact ventricular septum and tricuspid valve atresia.

Premature infants with patent ductus arteriosus (PDA) were included in the group with structural heart disease because they were assessed according to their need for pharmacological or surgical treatment, as was the case with the other structural cardiac defects seen during 1975.

A second group of neonates with nonstructural heart disease are reported separately (Rowe *et al.*, 1978).

Results

Table 1 summarises the final diagnoses in the neonates with structural congenital heart disease. The 10 most common defects are listed in order of frequency in 1975 and compared with the prevalence in 1965. The obvious change between the two years is the pronounced increase in the number of PDA in 1975. Although the absolute number of patients with D-transposition of the great arteries (complete transposition of the great arteries) rose by half, from 18 to 27, this malformation was still in second place in 1975.

Table 1 Incidence of common structural cardiac defects in neonates in 1965 and 1975

Final diagnosis	1965 (n=121)		1975 (n=226)	
	No.	%	No.	%
Patent ductus arteriosus	10	8	69	31
D-transposition of the great arteries (complete transposition)	18	15	27	12
Hypoplastic left heart syndrome	11	9	21	9
Ventricular septal defect	19	16	25	11
Coarctation of aorta syndrome	10	8	16	7
Hypoplastic right heart syndrome	7	6	8	4
Tetralogy of Fallot	10	8	9	4
Aortic valve stenosis	1	1	5	2
Dextrocardia	8	7	6	3
Pulmonary valve stenosis	3	2	6	3
Others	24	20	34	15

The sex ratio in both groups shows there were more boys than girls, but this trend is diminishing (Table 2). A significant reduction in the birthweight of the neonate with structural heart disease seen in 1975 is apparent (Table 2). In 1975, the mean age at first assessment was 6.8 days compared with 8.3 days in 1965 (Table 2). If the patients with PDA as the principal defect are excluded, the mean age at first assessment in 1975 was 5.4 days. The patients with PDA were mainly premature infants who did not show evidence of the shunt until the second week of life.

A greater reduction is noted in the interval from initial clinical assessment to cardiac catheterisation between 1965 (5.3 days) and 1975 (1.8 days) in Table 3. The two main indications for cardiac catheterisation in the neonate are central cyanosis and congestive heart failure (Rowe, 1970). With increasing severity of these indications, there is a greater urgency to proceed to cardiac catheterisation. This urgency is reflected in the shorter time interval to cardiac catheterisation in 1975 compared with 1965 (Table 4).

Table 2 Clinical data of neonates with structural cardiac defects assessed in 1965 and 1975

Year	No.	Sex ratio F/M	Birthweight (kg)		Age at first assessment (days)	
			Mean	SD	Mean	SD
1965	121	0.76	3.1	0.7	8.3	7.1
1975	226	0.84	2.8	1.0	6.8	2.2
			$t = 3.26$		$t = 2.26$	
			$P < 0.005$		$P < 0.025$	

Table 3 Interval from initial clinical assessment to cardiac catheterisation in neonates with structural heart disease, 1965 and 1975

Year	Mean interval (days)	No.
1965	5.3	66
1975	1.8	117
		$P < 0.001$

Table 4 The influence of cyanosis and congestive heart failure on the interval from initial clinical assessment to catheterisation, 1965 and 1975

Year	Cyanosis			Heart failure (mean, days)		
	Mild	Moderate	Severe	Mild	Moderate	Severe
1965	7.4	3.4	3.1	4.8	5.9	3.5
1975	1.3	0.2	0.2	4.3	2.6	0.3
	$P < 0.001$	$P < 0.001$	$P < 0.01$	NS	$P < 0.025$	NS

NS = not significant at conventional (5%) level.

Approximately one-half of patients with structural heart disease were admitted for noncardiac reasons, the majority being premature infants with PDA who initially were admitted for prematurity alone or with respiratory distress, jaundice, or hypoglycaemia. The remaining patients, who were admitted with suspected cardiac malformations, showed cyanosis or congestive heart failure. The severity of the defects was not based on the presence or absence of heart failure or cyanosis, but rather on the basis of the patient undergoing cardiac catheterisation, surgery, or dying, as was done in the New England series (Fyler *et al.*, 1972). The survival rates of babies with or without cyanosis and/or congestive heart failure were higher in 1975 than in 1965. Statistically, the higher survival rate in patients with congestive heart failure was not significant (Table 5). The presence or absence of the patients with PDA in the category with cyanosis and/or congestive heart failure had little effect as the survival rate was greater in 1975 than in 1965 (Table 6).

The assessment of the acid-base status of the 10 groups is best appreciated by their division into three physiological groups as in Table 7, the first with central cyanosis or hypoxaemia, the second with pressure load, and the third with volume load. Examination of blood pH shows that acidosis occurs in all severe cases, but not necessarily in the mild. Acidosis is more severe in left heart obstruction with impaired peripheral perfusion ($\text{pH} = 7.23$). The highest value of PCO_2 , hypercapnia, was in the volume load group (45 mmHg) and the lowest, hypocapnia, in the left heart obstruction group (25.2 mmHg). As expected, the maximum drop in PO_2 was in the hypoxaemia group (23.0 mmHg in D-transposition of the great arteries). The hyperoxic test indicates little rise in PO_2 in the hypoaemic group (23.0 to 24.7 mmHg mean).

In 1975, of 53 neonates undergoing cardiac surgery there were 37 (70%) survivors compared with a rate of 37% (13 of 35) in 1965 (Table 8). This increase in survivors undergoing surgery within the month in 1975 compared with 1965, was not due to increase in ligation of PDA. Despite the increase in this lesion, the number of ligations in 1975 in the first month of life was 5 with 4 surviving and 1 in 1965 who survived. The survival with Blalock-Hanlon

procedure in D-transposition of the great arteries (complete transposition), was much improved in 1975. 16 neonates underwent this procedure under one month without a death in 1975; only 3 of 12 survived in 1965. The results with this operation accounted for part of the major improvement.

Table 6 Survival of neonates with congenital structural heart defects (including and excluding patients with patent ductus arteriosus) and central cyanosis, congestive heart failure, or both, 1965 and 1975

Year	With PDA		Without PDA	
	Survivors	Total	Survivors	Total
1975	83 (62.9%)	132	70 (60.9%)	115
1965	33 (37.9%)	87	32 (37.6%)	85
	$\chi^2 = 11.35 \text{ P} < 0.001$		$\chi^2 = 9.63 \text{ P} < 0.005$	

Table 7 Arterial blood-gases measured in categories of congenital structural heart disease in the neonate, 1975

	pH	PCO_2 (mmHg)	PO_2 (mmHg)	Hyperoxic PO_2 (mmHg)
Hypoxaemia				
Severe (TGA)	7.30	36.9	23.0	24.7
Mild (T/F)	7.38	29.8	39.3	42.7
Pressure load				
Severe (HLH)	7.23	25.2	52.5	—
Mild (Co Ao)	7.25	39.7	59.9	—
Volume load				
Severe (Talner, 1970)*	7.35	45.0	56.0	—
Mild (VSD)	7.38	33.6	63.5	—

TGA = Transposition of great arteries; T/F = Tetralogy of Fallot; HLH = Hypoplastic left heart; Co Ao = Coarctation of aorta; VSD = Ventricular septal defect.

*29 volume loaded hearts in infants who were critically ill. 16 out of 29 had large ventricular communications.

Conversion: traditional units to SI— PCO_2 and PO_2 : 1 mmHg $\approx$ 0.133 kPa.

Table 8 Survival at least one month after surgery in congenital structural heart disease in the neonate, 1965 and 1975

Year	Total no.	Survival > 1 month	
		No.	%
1965	35	13	37.1
1975	53	37	69.8

$\chi^2 = 9.17$
 $\text{P} < 0.005$

Table 5 Survival of neonates with congenital structural heart defects and central cyanosis, congestive heart failure, or both, 1965 and 1975

Year	Cyanosis			Congestive heart failure			Combined cyanosis and congestive heart failure			
	Survived	Died	Total	Survived	Died	Total	Survived	Died	Total	Grand total
1975	45 (80.4%)	11	56	22 (59.5%)	15	37	16 (41%)	23	39	132
1965	21 (55.3%)	17	38	9 (39.1%)	14	23	3 (11.5%)	23	26	87
	$\chi^2 = 5.66 \text{ P} < 0.025$			$\chi^2 = 1.60 \text{ NS}$			$\chi^2 = 5.20 \text{ P} < 0.025$			

With medical treatment only, including patients with PDA, the survival rate in 1965 was 61% (54 out of 89) and in 1975 it was 75% (128 out of 170) ($\chi^2 = 5.3$). If the patients with PDA are excluded, the survival rate was 58% (46 out of 80) in 1965 and 67% (72 out of 107) ($\chi^2 = 1.8$) in 1975. The survival rate is higher in 1975 than in 1965, even with the PDA cases excluded, but the difference is not significant because of the smaller number of cases.

The management and prognosis of the neonate with heart disease may be influenced by the presence of extracardiac anomalies (Greenwood *et al.*, 1975). 26% of neonates with structural heart disease in 1975 had anomalies of systems other than the cardiovascular system. In 20% (45 out of 226) of the patients, admission was initiated because of the extracardiac anomalies. Mortality may have been influenced by the extracardiac anomalies in 28% (16 out of 57) with these additional defects.

Discussion

In 1960, the first report on congenital heart disease in the newborn from Toronto estimated the probable number of fatal cases there from the cases examined at the Hospital for Sick Children before death in the preceding 5 years. Fewer than half of these cases reached the hospital before death (Rowe and Cleary, 1960). In that period, 149 newborn babies with signs of heart disease survived the first month. The emphasis was on the difficulty of finding cases of congenital heart disease early and the need to separate severe from the less severe malformations. This was the preliminary to improving the survival of the seriously affected cases.

By 1965 the annual case load of structural heart disease was 121 patients. There were 13 survivors over a month postoperatively of 35 undergoing surgery in the neonatal period, a survival rate of 37%.

For comparison, a Buffalo survey for the period 1949–64, reported a survival rate of 31% (10 out of 32) in this age group (Lambert *et al.*, 1966). The figure from New Zealand for 1969–71 was 50% (6 out of 12) survival (Barratt-Boyes *et al.*, 1972).

Towards the latter part of the decade cardiac catheterisation was being widely used because of the improvement in mortality from heart disease at this age (Fyler, 1968, 1969; Varghese *et al.*, 1969). For example, the procedure of balloon atrial septostomy during cardiac catheterisation introduced in this period greatly accelerated the improvement in the survival rate of D-transposition of the great arteries. Surgical techniques suited to the smaller infant also became applicable (Mustard *et al.*, 1970; Edmunds *et al.*, 1972; Barratt-Boyes *et al.*, 1973).

The recognised need to improve case finding and

facilitate treatment was first appreciated by the New England Regional Infant Cardiac Program in 1969 (Fyler *et al.*, 1972) and within 4 years the case study within that programme had been shown to be complete (Nadas *et al.*, 1973).

At the turn of the decade, the Study Group of Congenital Heart Disease of the Intersociety Commission for Heart Disease Resources was recommending a stratified system to promote optimal care for all ages (Engle, 1972).

With better understanding of the biological changes occurring in hypoxia and congestive heart failure in the newborn with serious heart disease, management of these changes pre- and post-operatively improved and aided in the survival of the patients (Rowe, 1970; Talner and Ithuralde, 1970; Gersony and Hayes, 1972; Talner and Campbell, 1972).

In an attempt to evaluate the impact of these changes, we have reported here our experience for 1975 and compared it with that for 1965. The number of referrals and patients with structural heart disease (226) seen in one year, was equal to the total seen in the 5 years before the 1960 report. A few changes were noted in the incidence of various malformations. The obvious increase in the incidence of PDA is related to the survival of increasingly more premature infants in whom the incidence of PDA is rising (Siassi *et al.*, 1976).

The sick cardiac patient in 1975 was referred and seen earlier than in 1965. Before cardiac catheterisation, a number of newer noninvasive assessments helped to plan invasive investigations and decreased the time and improved the quality. Such assessment included the use of blood-gases to determine the acid-base status, the hyperoxic test for cyanotic patients, and echocardiography (Rowe, 1970; Talner and Ithuralde, 1970; Godman *et al.*, 1974; Hagler, 1976; Meyer, 1977; Williams and Tucker, 1977). The interval from first assessment to cardiac catheterisation decreased with greater severity of cyanosis or congestive heart failure, reflecting the trend towards earlier and more complete investigations.

With the more aggressive and newer approach to investigation, the prognosis with treatment is improving. In 1965, 61% of neonates were surviving one month after medical treatment, but by 1975 the figure was 75% ($0.02 < P < 0.025$). A number of factors influence the result of medical treatment. If the patients with PDA are excluded, the survival rate with medical treatment was 58% in 1965 and 67% in 1975 ($\chi^2 = 1.8$); the difference is not significant because of the smaller number of cases, particularly in 1975. Some patients will be treated medically because the lesion may be inoperable, as with aortic valve atresia, while others will

have quite minor defects and transient disturbances which respond well to medical treatment.

With improved surgery and perioperative care, the survival rate in the first month after surgery rose from 37% in 1965 to 70% in 1975 ($P < 0.005$).

Supported in part by grants from the Ontario Heart Foundation.

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Received 16 August 1978