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The Evolved Management of Neonates with Major Congenital Heart Disease

The survival rate of neonates with major congenital heart diseases is improving steadily

Congenital heart disease with major haemodynamic disturbances tends to present in the first year[1-5] and mostly the first weeks of life.[6,7] In the past, many of these heart lesions were considered inoperable and one-third to half of these symptomatic babies died within the first month of life.[5,8] Recent advances in the better understanding of the morphology, the introduction of non-invasive diagnostic modalities and improved surgical technique have all helped to enhance the survival of many of these sick newborns.[9] In this article, we will review the clinical profile of neonates with symptomatic congenital heart disease who were admitted into our hospital over the past decade. From the data collected, we will look at outcomes and relate this to the evolving management policies in our cardiac centre which is within the developing countries of Southeast Asia.

Study Population

Symptomatic neonates (≥ 30 days), presenting with tachypnoea, cyanosis, and/or heart failure who were admitted into the Grantham Hospital, University of Hong Kong, between January 1981 to December 1990 were included into the study. The hospital is a tertiary referral centre for neonatal heart problems for the whole territory of Hong Kong which has a population of 6 million.

Diagnosis of Congenital Heart Diseases

We utilised the sequential segmental approach[10] and achieved a definitive diagnosis of each neonate by either cardiac catheterisation, echocardiography,[11,12] surgery and/or autopsy findings. We included:

- All structural abnormalities of the heart or intra-thoracic great vessels that are of potentially functional significance
- Isolated persistent arterial duct which caused heart failure (but excluded it if it formed part of the presentation of persistent pulmonary hypertension of newborn)
- Neonates who had serious arrhythmia (with or without gross anatomical defects) and required medical treatment or pace-maker insertion.

The various congenital heart diseases were then grouped according to the major functional haemodynamic disturbances and the dominance of the heart lesions that demanded early treatment, e.g. when pulmonary atresia occurred in the setting of a complex heart lesion, the anomaly would be grouped under pulmonary outflow obstruction. Babies with coarctation of aorta with or without intracardiac shunts would be categorised into the subgroup of coarctation, under the major grouping of left ventricular outflow obstruction.

Method

The clinical profiles for each baby were reviewed from the hospital records and the following variables were included:

- age
- sex
- maturity and weight at birth
- year of admission
- ethnic group
- age of presentation
- symptomatology
- associated congenital anomalies
- method of definitive diagnosis before treatment (clinical, echocardiography, catheterisation)
- form of treatment received
- outcome for the neonatal admission and at follow-up.

Babies who survived a palliative neonatal surgery or medical treatment but who had significant haemodynamic disturbances were taken as 'at risk' when discharged from hospital. Neonates or babies who had corrective surgery performed at any stage and without major residual lesions were defined as 'well' on follow-up.

Usefulness of Echocardiography

A total of 782 babies (table I) with major congenital heart diseases were admitted over the 10 years. Based on the local birth rate,[13] and over 95% of the referrals from all over the terri-

TABLE I. Demographics of neonates with congenital heart diseases

	Number (%)
Sex	
Male	484 (62)
Female	298 (38)
Ethnics	
Chinese	760 (97)
Caucasians	22 (3)
Maturity	
Mature	676 (86)
Premature	106 (14)
Bodyweight	
> 2.5kg	628 (80)
< 2.5kg	154 (20)
Presentation	
1st week	642 (82)
2nd week	81 (10.5)
3rd week	26 (3)
4th week	33 (4.5)
Cyanosis	503 (64)
Heart failure	148 (19)
Tachypnoea	78 (10)
Rhythm disturbance	28 (4)
Associated anomalies (extracardiac)	102 (13)

tory were included, the incidence of neonates with symptomatic congenital heart disease was thus 1.02 per 1000 live births for the study period.

Since the introduction of echocardiography into the unit in late 1982, there has been an increasing trend to rely on the non-invasive investigation to arrive at a definitive diagnosis and implement interventional procedures (balloon septostomy) and surgical correction. Of the 466 neonatal procedures performed, 251 (54%) were based on echocardiographic findings alone (table II). The surgical mortality was 21% (98 of 466) while the overall neonatal mortality mounted to 30% (238 of 782). During the course of follow-up (15 months to 11 years), the cumulated total mortality was 44% (110 + 238) for the whole study group.

Pattern of Congenital Heart Diseases

The neonates were classified into 7 major groups with further subgroupings (table III). Pulmonary outflow obstruction occurred most frequently (n = 285, 36.5%) and correlated with the highest incidence of neonatal cyanosis in this study. Babies suffering from left ventricular outflow obstruction were second most common in occurrence (n = 173, 22%) and presented with heart failure with or without cyanosis. The group with left to right shunting (n = 119, 15%) and common mixing situation (n = 65, 8%) usually developed heart failure, while babies with transposition of great vessels (n = 96, 12%) presented with early cyanosis.

Group I – Pulmonary Outflow Obstruction

Among 285 babies, 158 had pulmonary atresia while 127 had valvar and/or subvalvar pulmonary stenosis. Prostaglandin infusion was instituted for babies who had a ductus-dependent pulmonary circulation. Various surgical procedures were performed on 177 neonates and the other 108 received medical treatment. With the exception of 2 major subgroups described below, a shunting operation (mostly modified Blalock-Taussig shunt) was the mainstay of initial surgical treatment for these neonates. An evolving surgical management was introduced for babies with pulmonary atresia and intact ventricular septum (n = 69).[14] In the early study period, 6 babies died when medical treatment alone was offered. Prior to 1984, indiscriminate right ventricular outflow reconstruction was performed for 14 neonates with 8 hospital deaths (57%) and 6 eventual long-term survivors. Since 1984, a staged operation with an initial surgery of either transventricular closed pulmonary valvotomy (n = 40) or a shunt operation (n = 9) was performed. The choice of operation was guided by the tripartite classification of the hypoplastic right ventricle of these neonates.[15,16] The subsequent procedures of either right ventricular outflow reconstruction (n = 9), balloon valvuloplasty (n = 11) for residual pulmonary stenosis[17] or a Fontan operation (n = 2) was performed at infancy or childhood. The neonatal hospital mortality of 29.5% (11/40) for the staged operation compared favourably ($p < 0.05$) to that of indiscriminate right ventricular outflow reconstruction.[14]

Similarly, for the subgroup of critical pulmonary stenosis with an intact ventricular septum (n = 26), initial right ventricular outflow reconstruction (n = 11) resulted in a high hospital mortality of 64% (9/11). Subsequent evolved management consisted of a staged operation of closed pulmonary valvotomy (n = 13) followed by either further right ventricular outflow reconstruction (n = 4) or balloon valvuloplasty (n = 3) for residual pulmonary stenosis, or a direct balloon valvuloplasty (n = 2) during the neonatal period. The evolved therapy greatly reduced the

TABLE II. Interventional procedures performed as guided by echocardiography

Types of interventional procedures	No. of cases
Balloon septostomy	53
Repair of coarctation[a]	50
Shunt operation	40
PAD ligation	29
PAD ligation + pulmonary arterial band	8
Closed pulmonary valvotomy (± shunt, ± septostomy)	29
Repair of TAPVC	17
Pulmonary arterial band	7
Arterial switch operation + septostomy	7
Others (e.g. balloon angioplasty)	11
Total	**251**

a ± ligation of PAD, ± pulmonary arterial band, ± septostomy.
Abbreviations: PAD = persistent arterial duct; TAPVC = total anomalous pulmonary venous connection.

TABLE III. Outcome of the neonates of various groups

	Study group [n = 782]	RVOT obstruction [n = 285]	LVOT obstruction [n = 173]	Left to right shunt [n = 119]	TGA complex [n = 96]	Common mixing [n = 65]	Miscellaneous [n = 44]
Neonatal outcome							
Discharged	544 (70%)	215 (75%)	73 (42%)	105 (88%)	89 (93%)	33 (52%)	29 (66%)
Died	238 (30%)	70 (25%)	100 (58%)	14 (12%)	7 (8%)	32 (48%)	15 (34%)
Latest follow-up							
Well	245 (33%)	62 (22%)	31 (18%)	78 (65%)	54 (56%)	17 (27%)	3 (7%)
'At risk'	122 (16%)	78 (27%)	13 (8%)	6 (5%)	9 (9%)	6 (9%)	10 (23%)
Lost	67 (9%)	30 (10%)	10 (5%)	13 (11%)	6 (7%)	2 (3%)	6 (13%)
Died	110 (14%)	45 (16%)	19 (11%)	8 (7%)	20 (21%)	8 (13%)	10 (23%)
Total mortality	348 (44%)	115 (40%)	119 (69%)	22 (19%)	27 (28%)	40 (62%)	25 (57%)

( ) = % for the specific group represented in the column.

hospital mortality to 20% (3/15), ($p < 0.005$).

Group II – Left Ventricular Outflow Obstruction

Of the 173 babies, 98 and 25 had coarctation and interruption of the aortic arch, respectively, 48 had hypoplastic left heart syndrome (mitral and aortic atresia) while 2 had severe aortic stenosis with a hypoplastic left heart. Conservative management was implemented for the babies with hypoplastic left heart (with aortic atresia or stenosis) and all 50 succumbed. Prostaglandin E_2 infusion was usually given to maintain patency of the arterial duct for babies with aortic arch obstruction (n = 123). Dopamine and ventilator supports were liberally instituted for most sick neonates. Medical treatment alone (n = 18) was offered when surgery could not be carried out because of the critical condition or rarely because of parental refusal. Otherwise, repair of the coarctation (n = 82) by subclavian flap aortoplasty or end-to-end anastomosis was carried out with ligation of the arterial duct (and banding of the pulmonary trunk when large intracardiac shunt existed).[18] Interruption of the aortic arch was repaired (n = 19) by either end-to-end anastomosis or interposition of a Goretex graft between the interrupted arch (together with ligation of the duct and banding of the pulmonary trunk).

The management for this group of sick babies was greatly facilitated by the use of echocardiography as the definitive diagnostic tool. The impact of such non-invasive techniques were most obvious with the subgroup of coarctation of the aorta. Amongst the 98 babies with aortic coarctation, 50 required catheterisation for diagnosis and eventually 22 died. As many as 15 babies succumbed soon after catheterisation without surgery. In contrast, 48 babies had echocardiography alone and subsequently 12 died ($p < 0.05$), only 6 of them were too ill to receive any operation after the non-invasive investigation ($p < 0.05$). The subsequent management on the long-term survivors with re-coarctation was greatly facilitated by the use of balloon angioplasty. Amongst the 64 survivors, 12 developed complications and 11 were successfully relieved of the aortic narrowing by therapeutic catheterisation.[19]

Left to Right Shunt

Persistent arterial duct (n = 51, 43%) and ventricular septal defect (48, 40%) were the most common lesions for this group. Of the total 119 babies, 67 were offered medical and 52 surgical treatment.

Echocardiography again offered a definite diagnosis for many babies. Investigation by either catheterisation or echocardiography, however, did not affect the neonatal survival for this group (17/20 *vs* 30/32, $p > 0.05$). The non-invasive technique was most useful for babies suffering from persistent arterial duct. Amongst the 51 neonates with persistent arterial duct, 41 were correctly identified but 1 baby had a false-positive diagnosis for coarctation of aorta. Medical treatment by fluid restriction, diuretics and indomethacin successfully closed off 18 ducts while 31 required surgical ligation. Two premature babies died before surgery as their conditions deteriorated rapidly on arrival.

Transposition of Great Vessels

Of the 96 neonates with simple transposition of great arteries and transposition with complex intracardiac anatomy, 92 underwent balloon septostomy but the procedure was not required in 4 cases who had an adequate atrial septal defect. Seven babies went for neonatal arterial switch operation and 69 other neonatal survivors underwent further surgery including Mustard's or Senning's operation with or without closure of a ventricular septal defect (n = 63), modified Blalock-Taussig

Shunt (n = 3), arterial switch operation (n = 1) and other complex surgery (n = 3).

The evolving management for this group was exemplified by the changing practice on neonates with simple transposition of great arteries (n = 55). During the early part of the study, all 19 babies had formal cardiac catheterisation followed by septostomy. Since 1983, septostomy was performed under echocardiographic guidance in the intensive care unit (n = 34).[20] Venous switch operations (n = 39) were then performed at around 1 year of age. The first 7 cases went for a Mustard's and the subsequent 32 babies a Senning's operation. The surgical mortality was 10% (4/39) but 7 other babies were lost to follow-up and were presumed dead prior to the venous switch operation. Thus the total mortality for babies destined for a venous switch operation was 28% (11/39). Since 1989, arterial switch operation was performed on 7 neonates with 2 hospital deaths (29%). The long-term survivors for the 2 different surgical groups were thus comparable (28/39 *vs* 5/7).

Common Mixing

The major subgroups for these babies included those with total anomalous pulmonary venous connection (n = 38), hearts with univentricular atrioventricular connection without any outflow obstruction (n = 14), common arterial trunk (n = 10) and double outlet right ventricle (n = 3).

Medical treatment alone was offered to 29 babies while corrective (as in total correction for anomalous pulmonary venous connection) and palliative (such as banding of the pulmonary trunk) operations were performed on 36 neonates. Eight of the 33 neonatal survivors underwent further corrective surgery (such as Fontan operation).

Miscellaneous

The miscellaneous group consisted of babies with arrhythmia, Ebstein's malformation of the tricuspid valve and other rare anomalies (e.g. cardiac tumour). Of the 44 neonates, 29 (66%) babies were alive at the latest follow-up.

Factors Enhancing Survival

Neonatologist and paediatricians looking after newborns are increasingly aware of the various early presentations of babies suffering from major congenital heart diseases. This is reflected by this retrospective review as most of the babies (92.5%) were referred to us within the first 2 weeks of life. A comparison on the frequency of occurrence of a cardiac anomaly between Western and our Oriental babies are reported elsewhere.[21] However, contrary to previous reports, coarctation of aorta is not rare amongst Chinese babies.[22-26] Many of the babies with left ventricular outflow obstruction presented in a 'collapsed' state and were successfully resuscitated by prostaglandin E_2 infusion with dopamine and ventilatory support in regional hospitals prior to transferal to our centre for management.

Our findings suggested that echocardiography as the definitive diagnostic tool has contributed significantly in lowering the mortality of neonates with coarctation of aorta. These babies, who were already extremely ill, often deteriorated further after catheterisation,[27] thus precluding them from surgical correction. Hence, overall mortality and mortality prior to surgery were higher for those who had catheterisation rather than echocardiography ($p < 0.05$) for the diagnosis.

Better understanding of the cardiac morphology of the hypoplastic right ventricle also enhanced the survival of neonates with pulmonary atresia and intact ventricular septum.[14] Instead of the indiscriminant approach of right ventricular outflow reconstruction, the evolved management of a staged biventricular repair for hearts with a tripartite right ventricle and an adequate size of the infundibulum, and a shunt operation followed by a univentricular repair (Fontan operation) for a diminutive right ventricle has significantly increased our neonatal and subsequent survival on follow-up.[14] The introduction of high torque guide wires and low profile balloon catheters also provided an alternative for management. Hence stenosis related to critical pulmonary valve stenosis,[28] residual pulmonary stenosis in babies after previous pulmonary valvotomy for pulmonary atresia[17] and critical pulmonary stenosis, and re-coarctation of the aorta[29] could all be alleviated by balloon dilation.

Improvement in intensive care and cardiac surgery has also revolutionalised the management for neonates with simple transposition of great vessels. Balloon septostomy could now be performed in our intensive care unit as guided by echocardiography.[23] The definition of the coronary arterial anatomy could be well assessed by the ultrasonic imaging.[30] The late introduction of the surgical technique in our hospital (since 1989) gave an apparent high mortality of 29% (2/7). Yet when the mortality for the venous switch operations and babies after septostomy who were lost while awaiting for this operation was put together, there was no statistical difference for the 2 surgical methods. In fact, the early neonatal and late mortality for our latest 36 babies who had arterial switch operation had dropped to 11% and 16%, respectively.[31]

Conservatism in Severe Cases

For our cohort of babies, the early neonatal mortality reaches 30% (298/782). This probably reflects the high risk associated with hearts with major haemodynamic disturbances presenting in this vulnerable interval. Amongst those who succumbed early, a considerable number of babies suffered from left ventricular

TABLE IV. Pattern of congenital heart diseases amongst neonates

Patterns	Number (%) [total = 782]
Pulmonary outflow obstruction	**285 (37)**
Stenosis	
TOF	37
Tetralogy complex + UVAC	13
Critical PS	39
+ Other intracardiac anomalies	26
	12
Atresia:	
+ IVS	69
+ VSD	36
+ UVAC	41
+ Other intracardiac anomalies	13
Left ventricular outflow obstruction	**173 (22)**
CoA complex	98
IAA	25
Hypo LV	48
AS	2
Left to right shunt	**119 (15)**
PAD	51
AVSD	11
Septal defects	54
Others	3
Transposition of great vessels	**96 (12)**
Simple TGA:	55
+ VSD	28
+ VSD + Mild PS	13
Common mixing	**65 (8)**
TAPVC	38
UVAC without outflow obstruction	14
Common arterial trunk	10
DORV	3
Others	**44 (6)**

Abbreviations: AS = aortic stenosis; AVSD = atrioventricular septal defect; CoA = coarctation of aorta; DORV = double outlet right ventricle; Hypo LV = hypoplastic left ventricle; IAA = interrupted aortic arch; IVS = intact ventricular septum; PAD = persistent arterial duct; PS = pulmonary stenosis; TAPVC = total anomalous pulmonary venous connection; TGA = transposition of great arteries; TOF = tetralogy of Fallot; UVAC = univentricular atrioventricular connection; VSD = ventricular septal defect.

outflow obstruction (100/238 = 42%). In particular, all 50 babies with hypoplastic left heart syndrome were managed conservatively without any attempt of either Norwood procedure or cardiac transplant. In this centre, which served a population of 6 million, we have a considerable waiting list for surgery and have focused our limited resources and manpower on cases that are more likely to achieve survival. We continue to adopt a conservative approach for the unfortunate group of neonates with hypoplastic left heart syndrome. As for babies who had pulmonary outflow obstruction and a complex heart with univentricular atrioventricular connection (often with atrial isomerism and anomalous pulmonary venous connection), many did not survive despite active palliative surgical correction. Their early death also contributed significantly to the total neonatal mortality for the present study. For those who survived the initial palliative shunt operation, many could eventually receive a Fontan procedure. Our latest data on survival for the Fontan operation are encouraging.[32]

Conclusion

The evolving management of neonates with major congenital heart disease, has greatly improved the survival of this group of patients born in our territory within the developing countries of Southeast Asia.

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