

A Precise of Present Surgical Treatment of Congenital Heart Disease

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It is usual to consider congenital heart disease in two groups;- Cyanotic and Acyanotic heart disease.

Cyanotic Heart Disease: Those cardiac structural abnormalities that result in some desaturated venous blood passing directly into the systemic arterial circulation without passing through the lungs. This type of patient is said to have a right-to-left shunt and exhibits central cyanosis. An example of this type of heart disease is "Tetralogy of Fallot".

Acyanotic Heart Disease: In this group the cardiac abnormality is not associated with a right-to-left shunt and therefore the patient is not cyanosed. There may be a left-to-right shunt i.e. part of the oxygenated blood that returns to the heart from the lungs passes back to the right side of the heart and is therefore "recirculated unnecessarily" through the lungs. An example of this type of heart disease is "Atrial Septal defect." Other congenital abnormalities may be associated with no shunts and therefore are also in the acyanotic group, example "congenital aortic stenosis."

It should be noted that certain congenital acyanotic heart abnormalities that have a left-to-right shunt, example "ventricular septal defect," "persistent ductus arteriosus; etc. if left unoperated, often develop progressive damage to the pulmonary arterioles due to the excessive blood flow. This causes a progressively rising pulmonary vascular resistance until finally the pulmonary vascular resistance is higher than the systemic vascular resistance. When this happens the left-to-right shunt (which has been getting smaller and smaller) finally reverses to become progressively cyanosed. This is termed "Eisenmenger Syndrome" and when this happens the condition is no longer operable and the patient is doomed to progressive cyanosis, breathlessness and finally death from hypoxia.

However, to clarify the present situation in congenital heart surgery I think it is better to consider the conditions in the following groups:

- I. Not operable.
- II. Ideally operable.
- III. Satisfactorily operable.
- IV. Unsatisfactorily operable.

I have divided them into these groups so that, in my opinion, it is possible to assess the validity of submitting a child to major heart operation with respect to operative risk and subsequent benefits and prognosis.

I. Not Operable

There are still a few congenital heart abnormalities that are so structurally severe that no operative structural or functional improvement can be performed. Examples of this are "mitral atresia" (absent mitral valve) and "left heart hypoplasia syndrome" (very small left ventricle, very small aortic valve and sometimes mitral valve, and hypoplastic ascending aorta). These types of abnormalities are not associated with a significant life expectancy and the new born baby usually fails to thrive in severe heart failure and dies within a few days or weeks.

As has been mentioned above some acyanotic heart abnormalities with left-to-right shunt (which are ideally operable in early years) may develop Eisenmenger Syndrome and become inoperable. This is an avoidable tragedy which is only due to delay in diagnosis or operation and should not occur.

II. Ideally Operable

This group of congenital abnormalities is ideally operable because surgery allows return to normal physiology of circulation and also normal or near

normal anatomy (these are the fundamental aims of cardiac surgery). Subsequent to operation the patient should have virtually normal life expectancy and exercise tolerance with no need to further operation later.

Congenital malformations within this group include (the respective operative risk and approximate unoperated life expectancy are appended in brackets):

Persistent ductus arteriosus	(0.2%:10-30 years)
Coarctation of Aorta	(0.5%:15-25 years)
Secundum Atrial Septal Defect	(0.5%:40-50 years)
Ventricular Septal Defect	(1.0%: 5-20 years)
Tetralogy of Fallot	(8.0%: 5-15 years)
Infundibular Stenosis with or without V.S.D.	(1.0%: 5-15 years)
Sinus Venosus Defect	(0.5%:40-50 years)
Pulmonary Valve Stenosis	(0.5%: 5-15 years)
Supra-aortic Stenosis	(1.0%: 5-15 years)
Sub-aortic Stenosis	(1.0%: 5-15 years)

The results of operation are so good with such relatively low risk that the failure to diagnose and operate on such patients could well be looked upon as medically negligent.

III. Satisfactorily Operable

Operation on this group of patients can produce marked improvement in their exercise tolerance and life expectancy. However although physiology of circulation is returned to virtual normality the anatomical improvement is less than ideal and later structural deterioration usually necessitates further heart operation later in life.

Congenital malformations within this group include (the respective operative risk and approximate unoperated life expectancy are appended in brackets):

Aortic Valve Stenosis	(1.0%: 5-10 years)
Primum A.S.D.	(3.0%: 5-20 years)
Pulmonary Atresia with central pulmonary trunk	(10%: few weeks)

The results of operation in this group are very satisfactory and despite the probability of requiring a second heart operation later in life the balance of advantages lie strongly on the side of operation which should be firmly advised.

IV. Unsatisfactorily Operable

In this group of abnormalities operation can produce good or partial correction of physiology of circulation but with only poor and usually non-durable anatomical improvement. The circulatory and exercise performance are improved but the long term outlook is probably poor. Therefore although life expectancy is prolonged it is probably nowhere near normal and also the operative risk is usually relatively greater.

Congenital malformations within this group include (the respective operative risk and approximate

unoperated life expectancy are appended in brackets):

Transposition of the Great Arteries	(12-15%: 5-15 years)
Truncus Arteriosus	(25%: few years)
Atrio-Ventricular Canal	(20%: few years)
Tricuspid Atresia	(15%:10-30 years)
Also all types of Univentricular Heart and common ventricle.	

Even if successfully operated upon these patients do not have a good long term prognosis. Although they are normally operated upon in all major cardiac surgical centres it is a delicate balance as whether it is medically sensible to do so. The parents are usually very keen to have anything done which may improve their child but viewing the whole clinical, social and prospective spectrum unemotionally and as scientifically as possible I am far from certain that cardiac surgery is totally beneficial. Each tragically affected child and parents must be carefully and individually assessed before advice is given.

Lastly in general terms the timing of operation is important. Whenever possible the operation is delayed until the child has reached a reasonable size (greater than 10kgs.) This makes the technical aspects of the operation easier and allows some co-operation from the child, (eg. atrial septal defect).

In many patients however, it is not advisable or possible to wait this long because of heart failure, failure to thrive or risk of irreversible damage to the pulmonary or systemic circulations eg. coarctation, persistent ductus arteriosus, ventricular septal defect, pulmonary stenosis, etc.



References:

- Beck, C.S. (1926), "Wounds of the heart: technique of suture" Arch. Surg., 13:205
 Segall, E.L. and Reid B.C. (1968): "The Heart and the law." (Macmillan, N.J.)
 Goidl, S.E. (1968) "Pathology of the Heart and Blood Vessels" (Charles C. Thomas).
 Hurst J. Willis Ed. (1982) "The Heart" (5th Edition)(Mc Graw Hill Book Co.).
 Chung E.K. Ed. (1980) "Cardiac Emergency Case" (2nd Ed.) (Lea & Febiger)
 Braunwald E (Ed) (1980) "Heart Disease" (W.B. Saunders).
 Willins F and Dry J. (1948) "A History of the Heart and Circulation." (W.B. Saunders)