

DR ASEEM R SRIVASTAVA

'Dr Aseem R Srivastava' is the Head for the Division of Pediatric Cardiothoracic Surgery at Artemis Hospital. He received his medical degree (MBBS) from GSVM Medical College, training in General Surgery (M.S) from King George Medical University, and training in Cardiothoracic Surgery (M.Ch) at GB Pant Hospital. He then worked as a Cardiac Surgeon in Delhi for about 4 years before leaving for the United States to work with few of the most prominent programs in Pediatric Cardiac Surgery.

He received his training in Pediatric Cardiac Surgery at 'The Children's Hospital of Philadelphia' and 'The Children's Hospital of Pittsburgh of UPMC' before joining The East Carolina Heart Institute where he gained extensive experience in Minimal Access Cardiac Surgery and Robotic Cardiac Surgery. On his return to India, Dr Srivastava joined Artemis Hospital in May 2015. Dr. Srivastava brings with him vast experience in reconstructive operations in neonates, infants and children and is a strong proponent of early corrective repair when possible.

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'A common misunderstanding is that the child needs to be of a particular age/ weight before cardiac surgery can be done- this is wrong and many babies either die without treatment or come late for surgery and are inoperable. With recent advances, cardiac surgery is safely possible even in newborn babies and Age or Weight do not matter in the treatment of Pediatric heart diseases'

SUGGESTED TIMING OF SURGERY IN CHILDREN WITH HEART DISEASES:

- **Atrial Septal Defect**

Ideal Age for **ASD closure 3-4 years**: this recommendation is based on the fact that a child with ASD is rarely symptomatic before the age of 4 years. If treated by this age the child is expected to lead a normal life with no sequel of the disease. Also, if treated by this age, lifelong memory attached to hospitalization and treatment does not form and the child will not remember anything associated with the heart condition apart from the scar.

A few complications of an ASD develop in late presenting children (Like Arrhythmias, Tricuspid regurgitation). These are permanent and related to cardiac enlargement and are not reversed by Surgery/Device Closure. And so, it is strongly recommended that surgery should be done before these develop.

Many children with this heart condition are diagnosed late, but even these children are operable and should receive treatment even if identified later in life.

- **Ventricular Septal Defect (VSD)**

Large VSD 'Do Not Close' on their own and so MUST be referred for Surgical VSD closure. Ideal Age for VSD closure: **6-12 months**
This recommendation is based on the following reasons:

1. Large VSD cause Pulmonary Artery Hypertension and many children, if left untreated, after the age of 12 months will turn INOPERABLE as pulmonary artery hypertension will become IRREVERSIBLE due to muscle hypertrophy in the pulmonary arteries.
2. Most symptoms of a Large VSD (Shortness of breath, repeated chest infections, difficulty in feeding) can usually be controlled by medications till about 3-6 months of age.

However, in a small proportion of children where symptoms cannot be controlled by medications VSD closure can be performed earlier.

Large VSD are unlikely to be operable after the age of 2 years.

Moderate Size VSD continue to be operable for a much longer time and should be referred when diagnosed even at a later age.

Small VSD should only be closed if they are causing Aortic Valve Prolapse or Aortic Regurgitation.

- **Patent Ductus Arteriosus (PDA)**

Ideal age for PDA closure is around **3 month** (earlier if symptoms cannot be controlled)

Children with PDA are less likely to become inoperable and so should be sent at any age for evaluation and treatment.

- **AV Canal Defect**

Ideal Age for Repair of an AV Canal Defect is **3-4 months**

This recommendation is based on the following reasons:

1. Complete AV Canal Defect will cause Pulmonary Artery Hypertension and many children, if left untreated, will turn INOPERABLE by the age of 6 months as pulmonary artery hypertension will become IRREVERSIBLE – due to muscle hypertrophy in the pulmonary arteries.
2. Most symptoms (Shortness of breath, repeated chest infections, difficulty in feeding) can usually be controlled by medications till about 3 months of age.

However, in a small proportion of children where symptoms cannot be controlled by medications, repair can be done earlier.

Babies with COMPLETE AV Canal Defect are unlikely to be operable after the age of 9-12 months.

In babies that have Downs syndrome in addition to an AV canal defect two things must be understood:

1. They tend to become inoperable earlier – by around 6-9 months of age
2. A significant proportion will also have hypothyroidism and should be tested and treated for it.

Children with PARTIAL AV Canal Defect (VSD is small and so PAH does not occur) continue to be operable for much longer time and should be referred when diagnosed, even at a later age.

- **Tetralogy of Fallot (TOF)**

The timing of surgery depends on 1. the child's saturations 2. the presence or absence of cyanotic spells and 3. on the natural history of untreated tetralogy of fallot.

Untreated: 25% die in the first year of life, 40% by 3 yrs., 70% by 10 yrs and this loss of life is almost always because of low saturations.

If the child's saturations are more than 75% and cyanotic spells are controlled with medications, then ideal age for a surgical repair is **6 -9 month**.

But if the child's saturations are less than 75% or the child is having repeated cyanotic spells that are not controlled with medications then surgery can be planned earlier.

Children with Tetralogy of Fallot that come late for treatment are almost always operable (although the risk of surgery increases with increasing age and this is because of the effects of long-standing low saturations on organ function). But since these children are operable - they should be sent for treatment even if identified later in life.

- **Transposition of Great Arteries (TGA)**

Timing of surgery is based on the presence or absence of a VSD in these children as this will define the natural history in case of untreated disease.

TGA without VSD - Survival without treatment – 80% at 1 week, 17% at 2 months, 4% at 1 year

Usual cause of death- Hypoxia

TGA with VSD – Survival without treatment 91% at 1 month, 43% at 5 months, 32% at 1 year

Usual Cause of Death- Heart failure

TGA without VSD: Surgical Repair is recommended at Diagnosis. Arterial switch operation is best done **within 2 weeks of life** but can be performed till 4-6 weeks of age. After this age, if the child survives, either a procedure to prepare the LV followed by Arterial Switch Operation (Staged surgery) is performed or a Senning's operation is done.

TGA with VSD: should undergo an Arterial Switch Operation with VSD closure **by 4-8 weeks of life**. But these children can usually be sent for treatment till about 6-9 months of age after which most will become inoperable.

- **Total Anomalous Pulmonary Venous Return (TAPVR)**

Natural History in untreated children: Untreated only 50% survive to 3 months and only 20% survive to one year of age

So surgical repair is recommended early in life- **as soon as the diagnosis of TAPVR is made**. Since these children do not become inoperable, even if the diagnosis is delayed, they should undergo surgical repair.

(Occasionally if the child is doing well, there is no obstruction to the flow of blood from the pulmonary veins to the left side of the heart, then under close follow up, surgery may be delayed till 3 months of age)

- **Coarctation of Aorta**

Natural History of Untreated disease:

30% risk of death in the 1st year of life (acute heart failure),
1.6 % per year thereafter (aortic rupture, endocarditis,
intracranial hemorrhage)

So surgical repair is recommended early in life- **as soon as the diagnosis of Coarctation of Aorta is made**. Since these children do not become inoperable, even if the diagnosis is delayed, they should undergo surgical/ Interventional treatment.

- **Truncus Arteriosus**

Natural History in untreated children: Only about 50% survive beyond the 1st month, 30% beyond 3 months, 15% beyond 6 months and only 10% survive beyond 1 year.

And most children that survive beyond 6- 9 months of age will be INOPERABLE as pulmonary artery hypertension would be IRREVERSIBLE because of muscle hypertrophy in the pulmonary arteries.

So surgical repair is recommended **early in life at Diagnosis (ideally before 3 months, but in cases of late presentation children up to 9 months of age may be sent)**

- **Single ventricle defects**

Staged treatment starts at diagnosis (ideally within three month of life)

Depending on whether the child has Pulmonary artery Hypertension versus Pulmonary stenosis (narrowing in the pulmonary artery), stage one will differ (PA Band Versus BT Shunt) but Stage 2 and 3 remain the same under both situations.

These children will usually need three operations:

Stage 1- PA Band operation 2-3 months of age (in Pulmonary artery hypertension)

Babies with Pulmonary artery hypertension tend to become inoperable by the age of 5-6 months, if left untreated.

OR

BT Shunt operation- 2-3 months of age (in Pulmonary artery stenosis- if saturations are less than 75%). Since these children have pulmonary stenosis, they do not become inoperable, even if the diagnosis is delayed and should undergo surgical repair.

Stage 2- BD Glenn operation at 6-9 months of age

Stage 3- Fontan operation- 4-5 years of age

- **Pulmonary atresia**

These are a complex subset of heart conditions- usually dependent on the continued patency of PDA which is very unpredictable and so- treatment starts at Diagnosis



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