Adult with Cyanotic Congenital Heart Disease

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Cyanotic Adult
Adult Cyanotic Congenital Heart Disease

Population Sub Types

- Operated cases after total correction
- Those who have had palliative surgeries to prolong life and improve physical capacity
  - Glenn’s & Fontan’s
  - Aorto pulmonary Shunts
  - Physiological Correction for the dTGA – Senning’s
- Those in whom no definite treatment is available only supportive therapy given
- Late presenters
Cyanotic Congenital Heart Disease in Adults

Cyanosis

Increased Erythropoiesis

Polycythemia

- Hyper viscosity
- Platelet dysfunction
- Deranged Coagulation parameters
- Fibrinolysis
- Iron deficiency

Contd....
Cyanotic Congenital Heart Disease in Adults

Hyper viscosity

- Headache
- Lethargy
- Confusion
- Visual disturbances
- Easy bruising, epistaxis, gingival hypertrophy
- Traumatic and per-operative bleeding
Cyanotic Congenital Heart Disease in Adults

Cyanosis

- With reduced pulmonary blood flow
  - Collaterals develop may cause
    - Haemoptysis, per-operative bleeding

- Thrombo-Embolic Complications
  - CVA
  - Pulmonary Hypertension
  - Brain Abscess

- Dizziness, Fatigue
- Endocarditis
- Hypoxic Cardiomyopathy

Contd....
TOF with Hypoplastic LPA & Blocked BT Shunt

Showing Rib notching on left side
Intercostal Collaterals

Dilated tortuous LMCA and Collaterals
50 yrs TOF 70 kg

30 yrs Executive, TOF repair with TV and AV replacement (Endocarditis)

40 yrs TOF with CABG

74 yrs Infundibular Stenosis with right to left atrial shunt
Cyanotic Congenital Heart Disease in Adults

Cyanosis

- Hyperuricemia
- Renal dysfunction
- Arthropathy
Adult Cyanotic CHD

Other Modes of Presentation

- Heart Failure
- Arrhythmias
- Skeletal abnormalities
- Sudden death
Non cardiac sequelae of Polycythaemia

- Hyperuricemia
  - Gout, Urate nephropathy

- Glomerular sclerosis
  - Manifests initially as proteinuria

- Hypertrophic Osteoarthropathy

- Gall stones and Cholecystitis
Cyanotic Congenital Heart Disease in Adults

Psychological Issues

- Subclinical distress
- Psychiatric disorders
- Abnormal neuro cognitive function
  - Chronic illness and disability
  - Hypoxia
  - Ischemia
  - Surgeries
  - Parental over protection
Adult Cyanotic Heart Disease

Special Challenges:

- Employment
- Interpersonal relations
- Psycho social issues
- Assuming responsibility of their own health care
- Sports participation
- Family Planning
- Uninsurable
Cyanotic Congenital Heart Disease

Cyanosis

General measures

- Avoid dehydration
- Avoid areas of hypoxia
  - High Altitude
  - Crowded places
- Avoid severe physical exertion
- Strict contraceptive measures in female patients
- SBE prophylaxis
- Iron, Folic acid supplement
- Monitor for Polycythemia

Contd....
Cyanotic Congenital Heart Disease in Adults

General Measures

- Hb > 18 Gm %
- PCV > 65
- With symptoms of hyperviscosity

$$\Rightarrow$$ Judicious Venesection
- Remove 500 ml blood
- With isovolumic replacement
- Repeat if necessary

- Intraoperative Bleeding

$$\Rightarrow$$ Platelets, Aprotinin
Cyanotic Congenital Heart Disease in Adults

Tetralogy of Fallot’s physiology

Can be offered total or palliative correction if anatomy favorable
Adults with Congenital Heart Disease

Tetralogy of Fallot physiology

Good intra cardiac mixing and pulmonary stenosis

- Tetralogy of Fallot’s
- Double outlet right ventricle
- Tricuspid atresia
- D or L transposition of great arteries
- AVSD with pulmonary stenosis
- Univentricular heart
Complex CHD after Fontan procedure – Special Issues

- Best option for patients who cannot have two ventricular repair
- Problems faced in long term
  - High central venous pressure
  - Low cardiac out
  - \(\uparrow\) Systemic vascular resistance
  - Heart failure – common with single RV
  - AV valve regurgitation
  - PLE

Contd.....
Complex CHD after Fontan procedure

- Thrombo Embolism
- Arrhythmias
- Need for pacing
- Right to left shunt across fenestration
- Venous collaterals
- Plastic Bronchitis
- Sudden death

Contd.....
Psycho social issues

- Life long anti platelet or anticoagulants
- Sluggish venous circulation
- Low cardiac output in relation to activity
Problems after Fontan Operation

- Persistent post operative pleural effusion
- Hospital stay > 18 days poor outcome
- Renal dysfunction
- Hepatic dysfunction
- May need CT/MRI
Adults with Cyanotic CHD - Operated

Post Arterial Switch for dTGA

- Yearly Follow up with EKG and Echo
- Coronary arterial obstruction
- Aortic root dilation, Aortic regurgitation
- Peripheral pulmonary stenosis
- Need CT Angio and thallium perfusion study for coronary issues after 10 years electively, early if any symptoms
Adults with Cyanotic Congenital Heart Disease

Post Senning operation for dTGA

- RV dysfunction – RV systemic ventricle
- Tricuspid regurgitation
- Atrial arrhythmias
- Baffle obstruction
- Need yearly follow up with EKG and Echo
Cyanotic CHD in Adults

- After Corrective Surgery
  - TOF physiology – Total Correction
  - Rastelli Operation
    → DORV, VSD, PS
    → Pulmonary atresia with VSD
    → dTGA, VSD with PS

- Need life long yearly follow up with Echo. & EKG
  - To detect need for medications
  - Detect arrhythmias
  - Need for reoperation
  - MRI ±
Adult with Cyanotic CHD Operation

Conduit Operation

- Persistant Trunkus Arteriosus
- TOF with absent pulmonary valve
- TGA VSD with PS

- Follow up 6 months – 1 year
- Conduit Obstruction
- Effects of Pulmonary regurgitation
Adults with Operated CCHD

TOF and DORV after Corrective Surgery

Severe pulmonary regurgitation mostly with TAP

- Progressive right ventricular dilation
- Right ventricular systolic and diastolic dysfunction
- Need for pulmonary valve replacement
- Arrythmias
- Sudden Death

TOF, physiology – palliated

Those with inadequate PA anatomy

After Shunt Surgery

- PAH
- High incidence of endocarditis
Ebstein’s Anomaly of Tricuspid Valve

- Severe displacement of proximal attachment of Tricuspid valve
  - CHF
  - Tricuspid regurgitation
  - RA and RV enlargement
  - Right to left atrial shunt
  - Arrhythmias
  - WPW Syndrome

Management

- Tricuspid valve repair/replacement
- EP Study – RF ablation if needed
Adults with Cyanotic Heart Disease

Lesions at High Risk for Endocarditis

- Unrepaired patients
- Palliated with Shunt/Conduits
- Prosthetic material used at surgery
- Previous Endocarditis
Pregnancy with Cyanotic Congenital Heart Disease

➢ Pregnant women with cyanotic CHD should be managed by the patient's obstetrician and cardiologist concurrently

➢ Post partum the patients may need ICU monitoring even in relatively minor procedure or uncomplicated deliveries, as they are prone to severe bleeding

➢ Risk for Foetus

  - Abortion
  
  - Fetal Death

Contd.....
Adult with Cyanotic CHD

Pregnancy

- Not well tolerated in patients with cyanotic CHD
  - Fetal loss
  - Maternal Death
Adult Cyanotic CHD - Presentation

Inoperable lesions

- Shunt lesions with pulmonary vascular disease
- Admixture lesions with pulmonary vascular disease
- TOF physiology with poor pulmonary anatomy
Eisenmenger’ Syndrome

- Severe PAH with right to left shunt due to ↑ PVR secondary to
  - ↑ PBF acyanotic shunt lesion
  - ↑ PBF with Cyanotic CHD

- Severe Hypoxia, exertional dyspnea, chest pain, syncopal attacks, haemoptysis, headache, giddiness, arrhythmias, sudden death

- Strict contraceptive measures in female patients

- No specific therapy is effective

- Heart lung transplant
Adult Cyanotic Congenital Heart Disease

- Increasing population of operated CCHD patients
- Continue to see unoperated adult patients with CCHD challenging to pediatric cardiac surgeons
- Need to deal with unusual situations like TOF physiology beyond 40 years
- Rarely operable admixture lesions like TAPVC without obst.
- Ebstein's disease presenting late – Operable
- Close cooperation with adult cardiologist and surgeon
- Develop GUCH programme as proposed by Dr. Jane Somerville
Thank You