Complete Summary

GUIDELINE TITLE

Management of grown up congenital heart disease.

BIBLIOGRAPHIC SOURCE(S)

Deanfield J, Thaulow E, Warnes C, Webb G, Kolbel F, Hoffman A, Sorenson K, Kaemmer H, Thilen U, Bink-Boelkens M, Iserin L, Daliento L, Silove E, Redington A, Vouhe P, Priori S, Alonso MA, Blanc JJ, Budaj A, Cowie M, et al. Management of grown up congenital heart disease. Eur Heart J 2003 Jun;24(11):1035-84. [82 references] PubMed

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Grown up congenital heart disease

GUIDELINE CATEGORY

Diagnosis
Evaluation
Management
Treatment

CLINICAL SPECIALTY

Anesthesiology
Cardiology
Critical Care
Emergency Medicine
Family Practice
Internal Medicine
Medical Genetics
Nursing
Obstetrics and Gynecology
Pediatrics
Psychiatry
Psychology
Radiology
Surgery

INTENDED USERS

Advanced Practice Nurses
Clinical Laboratory Personnel
Physicians
Psychologists/Non-physician Behavioral Health Clinicians

GUIDELINE OBJECTIVE(S)

- To evaluate provisions for care for grown-ups with congenital heart disease in Europe
- To make recommendations for improvement in organization facilities, training, and research
- To promote collaboration between the various professional groups involved in the care of adolescents and adults with congenital heart disease, administrators, and those who provide resources for healthcare

TARGET POPULATION

Adolescents and adults with congenital heart disease

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnostic Investigations

1. Electrocardiography
2. Chest x-ray
3. Echocardiography
4. Transoesophageal echocardiography
5. Cardiac catheterization
6. Magnetic resonance imaging
7. Holter monitor
8. Exercise function
9. Stress test
10. Electrophysiological study
11. Echo-Doppler

12. Radionucleotide studies
13. Invasive studies
14. Cardiac catheterization
15. Computed tomography
16. Signs and symptoms

Treatment/Management

1. Surgery
2. Device closure
3. Catheter closure in muscular ventricular septal defects (VSDs)
4. Valve repair or replacement
5. Closure of re-/residual atrial septal defect (ASD)
6. Atrioventricular (AV) universal pacemaker
7. Balloon valvuloplasty
8. Surgery with ablation
9. Balloon dilatation/stenting
10. Radio frequency ablation catheter intervention for pulmonary valve insertion
11. Surgical replacement of conduits
12. Mechanical valve replacement
13. Prosthesis
14. Homograft
15. Conversion to total cavopulmonary connection (TCPC)
16. Transplantation
17. Closure of fenestration
18. Atrioventricular sequential pacing
19. Valve-sparing operation
20. Transcatheter closure
21. Tricuspid valve replacement
22. Conversion to arterial switch (pulmonary artery banding)
23. "Double switch"

MAJOR OUTCOMES CONSIDERED

- Left ventricular function
- Haemodynamic changes
- Morbidity and mortality related to congenital heart disease and its complications
- Quality of life
- Survival
- Intellectual and educational attainment
- Recurrence risk
- Ability to participate in sport/physical activity

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

A. The data were derived from multiple randomized clinical trials.
B. The data are based on a limited number of randomized trials, nonrandomized studies, or observational registries
C. Consensus opinion of the experts

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

The committee reviewed and ranked the evidence supporting the current recommendations according to the strength of evidence against or in favour of a particular treatment or diagnostic procedure (see "Rating Scheme for the Strength of the Evidence" field).

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

In controversial areas, or on issues without evidence other than usual clinical practice, a consensus was achieved by agreement in the expert panel after thorough deliberations.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Class of Recommendation

Class I: Conditions for which there is evidence or general agreement that a give procedure or treatment is useful and effective.

Class II: Conditions for which there is conflicting evidence or a divergence of opinion about the usefulness/efficacy of a procedure or treatment.

Class IIa: Weight of evidence /opinion is in favour or usefulness/efficacy

Class IIb: Usefulness/efficacy is less well established by evidence/opinion.

Class III: Conditions for which there is evidence and/or general agreement that the procedure/treatment is not useful/effective and in some cases may be harmful.

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The document was reviewed by the members of the Committee for Practice Guidelines & Policy Conferences (CPGPC,) which also decided whether the document needed to be reviewed by external reviewers and by European Society of Cardiology (ESC) Board Members.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Transition from paediatric to adult care

- Paediatric cardiologists should begin to inform patients and families regarding transition from around the age of 12, with a flexible policy of transition at age 14 to 16 years. Subsequent transfer to the adult service can again occur at a flexible age of approximately 18 years. Each paediatric cardiac unit should establish a coordinated process to link with a specialist centre for grown-ups with congenital heart disease.
- The patient and his or her family will have developed a firm bond with the paediatric cardiologist over many years, and it is desirable that the paediatric cardiologist is involved in the transition service, together with the adult specialist(s).
- The transition clinic requires input from administration and other healthcare professionals. The nurse specialist is a key person and should be experienced in counselling of adolescents and their families and should be responsible for coordinating transfer arrangements.
- The patient and his or her family should be given a detailed written plan in advance of handover. This should include key information about treatment in

childhood (such as previous investigations and operations). It is part of an education process, which must be tailored to the varying levels of maturity and intellect of individual patients. The adolescent and his or her family need to understand their cardiac condition, healthcare needs, and prognosis. In particular, they should know about their medication, possible side effects and interaction with other drugs (including alcohol), and they should be fully informed about endocarditis prophylaxis. They also require guidance concerning exercise, contraception, pregnancy, career planning, travel, and insurance. They must be properly informed of possible future complications of their condition and likely associated symptoms. They must know how to operate within the adult healthcare system in order to obtain appropriate medical advice; both locally and when they are away from home (during studies, occupation, and travel). This process must be handled sensitively and cannot occur during a single consultation. The patient should be able to have private discussions, not only with the cardiologist, but also with the nurse specialist. Many children reach adolescence with little understanding of the implications of their condition, due to parental overprotection and lack of direct discussion. Parents often find it difficult to let go of their offspring and may need support to allow the adolescent to become independent. Time should be allocated to discuss sensitive issues such as contraception, family planning, pregnancy, recurrence risk, and sport.

- The carefully considered plan of medical management (including follow-up) should be based on the patient´s condition and prognosis, education and availability of local medical services. Unit protocols are very useful and the plan needs to be communicated to the primary care physician and other doctors involved (e.g. university healthcare services). Shared care with local physicians is appropriate for many patients. This is particularly important in special circumstances, such as noncardiac surgery or emergencies. Close liaison and good communication needs to be established at the transition stage.
- Transition of care should be a gradual process both for the patient and the medical practitioners. It is essential not to bombard the patient with an overwhelming amount of information, which can induce denial and lack of attendance. There should be continued opportunities for joint discussion between paediatric and adult specialists (both medical and surgical) and there should be feedback from both the transition and adult clinics to the paediatric cardiology team.

Training of practitioners in grown-up congenital heart disease

Required knowledge and skills

- Expertise of congenital heart malformations and management in infancy and childhood
- Expertise in general medicine and noncardiac diagnosis in adults
- Expertise in adult cardiology including coronary artery disease management
- Skill in the following procedures in adults with special reference to congenital heart disease:
    - Echocardiography (including transoesophageal)
    - Cardiac catheterization
    - Pacing and electrophysiology
    - Postoperative care

- Understanding of the physiological changes of pregnancy
- Understanding of the psychosocial aspects of adolescence
- Experience of life style counselling for adolescents and adults with congenital heart disease
- Expertise in clinical research methodology

Trainees from paediatric cardiology

- Three years in general paediatric cardiology
- 6 to 12 months in general medicine and adult cardiology
- At least 12 to 18 months in a specialist grown-up congenital heart disease centre
- For those aiming at an academic career in grown-up congenital heart disease, an additional year in research or specialist training is required.

Specific requirements for training (from pediatric cardiology) are familiarity with:

- Common adult cardiac problems, such as ischaemic heart disease, hypertension, arrhythmias and their treatment
- Coronary angiography (there is no need to learn coronary angioplasty)
- Recognition and management of arrhythmias related to operations for congenital heart disease
- Management of pacemakers and practical experience in their insertion. Management of implanted catheter defibrillators
- Management of middle-aged and elderly patients
- Problems of pregnancy in relation to cardiac problems, and effects of drugs on the patient and foetus
- Understanding the indications and contraindications of various methods of contraception
- Advice about exercise in various congenital heart abnormalities, both unoperated and operated, and what limitations, if any, apply
- Advice concerning driving of motor vehicles
- Information about life insurance and mortgages for patients with various unoperated and operated forms of congenital heart disease
- Vocational advice

Trainees from adult cardiology

- Three years in general cardiology including general medicine
- One year of paediatric cardiology in a centre with an active surgical and catheter intervention programme
- At least 12 to 18 months of training in a specialist grown-up congenital heart disease (GUCH) centre for those aiming at an academic career in grown-up congenital heart disease
- Additional year in research or specialist training is required.

Specific requirements for training (from adult cardiology) are:

- Foundation courses on congenital heart malformations and echocardiography
- Experience of echocardiography in infants and children, preoperatively and postoperatively. At least 250 echocardiographic examinations should be performed and an additional 25 transoesophageal echocardiograms.

- Experience of cardiac catheterization and angiography of common congenital heart anomalies. A minimum of 100 procedures should be performed independently.
- Familiarity with interventional procedures in congenital heart disease with participation in at least 25 procedures
- Involvement in management of and responsibility for at least 300 patients with congenital heart disease. These should include patients in the early postoperative period and those assessed during long-term follow-up.
- Attendance at weekly conferences of paediatric cardiologists and cardiac surgeons and participation in all of the teaching activities of the department
- Knowledge of genetic implications and familiarity with genetic counselling
- Understanding of psychosocial problems of adolescence, including schooling, bullying, and other behavioural issues, such as sex and drugs
- Vocational advice
- Problems of pregnancy in congenital heart disease
- Contraception advice for congenital heart disease

Specific surgical challenges

Preservation of myocardial function

- Aortic cross clamping should be avoided where possible. Most operations involving the right heart (e.g., replacement of right ventricle to pulmonary artery valved conduit or extracardiac Fontan procedure) can be carried out under normothermic cardiopulmonary bypass with mono- or bicaval cannulation and a beating heart.
- When aortic cross clamping is necessary, cross-clamp time should be kept as short as possible, and particular attention should be paid to cardioplegic myocardial preservation.

Strategies for myocardial protection and cardioplegia

- Use of the appropriate cardioplegic solution
- Induction of myocardial hypothermia using a cold blood cardioplegic solution
- Maintenance of diastolic arrest and hypothermia using multi-dose blood cardioplegia. This is particularly important in cyanotic adults, in whom noncoronary collateral vessels to the heart may result in wash out of cardioplegia and myocardial rewarming.
- Enhanced warm blood reperfusion administered prior to aortic unclamping under careful pressure monitoring
- Adequate venting of the heart to avoid ventricular distention, wall tension increase, and subsequent inadequate delivery of cardioplegic solution

Atrial septal defect (ASD)

<u>Introduction and background</u>

- Common defect which may be diagnosed first in adult life

<u>Survival/adult life</u>

- Small defects: excellent prognosis
- Large defects: reduced survival, depending on age at treatment

Haemodynamic issues

- Pulmonary hypertension
- Right ventricle (RV) dilation/failure
- Potential for paradoxical embolism
- Reduced left ventricle (LV) compliance

Arrhythmia/pacing

- Atrial arrhythmia (atrial fibrillation and flutter)
- Sick sinus syndrome
- Pacing rarely required

Investigations

- Electrocardiography (ECG)
    - Baseline: if clinically indicated (arrhythmias)
- Chest x-ray
    - Baseline: otherwise little value
- Echocardiography (ECHO)/transoesophageal echocardiography (TOE)
    - Baseline: location, size, RV size, pulmonary arterial (PA) pressure, Qp:Qs, associated lesions
    - TOE usually performed in older patients and at device closure
- Catheterization
    - Device closure
    - Peripheral vascular resistance (PVR) assessment
- Magnetic resonance imaging (MRI)
    - Rarely helpful
- Holter monitor
    - If symptomatic arrhythmia
- Exercise function
    - Baseline: little value

Indications for intervention

- Large defects (>10 mm) unless pulmonary vascular disease (peripheral vascular resistance >8 Um^2; L-R shunt <1.5; no response to pulmonary vasodilators)
- Paradoxical embolism

Interventional options

- Surgery or device closure (stretched diameter <38 mm)

Posttreatment outcome

- Low risk procedure unless pulmonary vascular disease (PVD)
- Late intervention less successful

Endocarditis

- Very rare
- Prophylaxis not indicated

Pregnancy/contraception/recurrence/fetal

- No contraindications unless PVD
- No restrictions for contraception
- Consider fetal ECHO

Recurrence/genetics

- 3% of first degree relatives
- Familial ASD (with long PR interval)
- Autosomal dominant

Syndromes

- Holt Oram: upper limb deformity
- Autosomal dominant

Sport/physical activity

- No restrictions unless moderate/severe PVD

Insurance

- Category 1
- Generally no problem if defect closed early

Follow-up interval

- Early repair (<30 years): no problems; discharge
- Late repair: regular follow-up

Follow-up care

- Level 2

Unresolved issues

- Surgery vs. device closure
- When to close in pulmonary hypertension (PHT)
- Concomitant Maze procedure
- Upper age limit for surgery
- Patent foramen ovale closure in patients with suspected paradoxical embolism

Ventricular septal defect (VSD) – unrepaired

Introduction and background

- Significant ventricular septal defects usually repaired in childhood
- See ventricular septal defect and PVD (Eisenmenger) but diminishing
- Small ventricular septal defect or postoperative septal defect common in adults
- Eisenmenger patients becoming less frequent

Survival/adult life

- Excellent for small ventricular septal defect
- Large ventricular septal defect may have pulmonary vascular disease (Eisenmenger)
- May develop aortic regurgitation

Haemodynamic issues

- Left-right shunt
- LV dilatation and impaired function
- Aortic regurgitation
- Pulmonary vascular resistance in uncorrected large ventricular septal defect

Arrhythmia/pacing

- Rare

Investigations

- Chest x-ray
    - Baseline: cardiomegaly
- ECG
    - Routine
    - Rhythm chamber enlargement
- ECHO
    - Number size and location of defects
    - LV/RV function
    - Aortic regurgitation
- TOE
    - If transthoracic echocardiography (TTE) image inadequate
- Catheter
    - Pulmonary vascular resistance
    - Associated lesions
- MRI
    - Rarely helpful
- Holter monitor
    - Only if symptomatic
- Exercise test
    - Only if symptomatic
    - Sports counseling

Indications for intervention

- Left-right shunt with left heart volume overload
- Reversible pulmonary hypertension
- Aortic regurgitation
- Associated abnormalities (RV outflow tract, subaortic stenosis)
- Previous endocarditis

Interventional options

- Surgery
- Catheter closure in muscular VSD(s)

Posttreatment outcome

- Good surgical results

Endocarditis

- Prophylaxis in all

Pregnancy/contraception

- No contraindications in uncomplicated VSD
- Pregnancy contraindicated in pulmonary vascular disease (Eisenmenger disease)

Recurrence/genetics

- Occasionally familial
- Usual recurrence risk
- Common cardiac anomaly in syndromes (e.g., Down´s)

Sport/physical activity

- No restriction in small VSDs

Insurance

- Small VSDs category 1

Follow-up interval

- Infrequent follow-up unless haemodynamic abnormalities (e.g., aortic regurgitation)

Follow-up care

- Small ventricular septal defect 3; pulmonary vascular disease (Eisenmenger) 2; aortic regurgitation/complicated haemodynamics 1

Unresolved issues

- Optimal management of Eisenmenger patients

Repaired ventricular septal defect

Introduction and background

- Common lesion
- Most patients now adults

Survival: adult life

- Excellent survival
- Occasional residual shunt
- Some develop RV or LV outflow tract obstruction
- Some develop aortic regurgitation

Haemodynamic issues

- Residual shunt
- Ventricular function
- Aortic regurgitation
- New haemodynamic abnormalities (RV outflow obstruction)

Arrhythmia/pacing

- Rare atrioventricular (AV) block, ventricular arrhythmia

Investigations

- Chest x-ray
    - Baseline: cardiomegaly
- ECG
    - Rhythm
- Echo
    - Residual VSD(s)
    - LV/RV function
    - Aortic regurgitation
- TOE if TTE insufficient
    - TOE only if TTE inadequate
- Catheter
    - Rarely required
- MRI
    - Rarely helpful
- Holter monitor
    - Only if symptomatic
- Stress test
    - Only if symptomatic
    - Sports counselling

Indications for intervention

- If residual VSD; see "unrepaired VSD"

Interventional options

- See "unrepaired VSD"

Posttreatment outcome

- See "unrepaired VSD"

Endocarditis

- Prophylaxis if residual VSD
- Questionable in closed VSD

Pregnancy/contraception

- No contraindications in uncomplicated closed VSD
- Pregnancy contraindicated in PVD (Eisenmenger)

Recurrence/genetics

- See: "unrepaired VSD"

Sport/physical activity

- No restriction in closed VSD

Insurance

- Category 1

Follow-up interval

- Can discharge if closed VSD without any residual abnormalities
- Infrequent follow-up for minor residual lesions

Follow-up care

- Eisenmenger 2; small VSD 3; aortic regurgitation/complicated haemodynamic 1

Unresolved issues

- None

Postoperative complete atrioventricular septal defect (AVSD)

Survival/adult life

- Unoperated survivors develop PVD.
- Surgical results markedly improved
- Status after repair depends mostly on left AV valve function
- Many patients have Down´s syndrome

Hemodynamic issues

- Left AV-valve regurgitation (± stenosis)
- PVD
- Late subaortic stenosis

Arrhythmia/pacing

- Risk of complete heart block low (<2%)
- Atrial arrhythmias, especially with left AV-valve dysfunction

Investigations

- Chest x-ray
    - Cardiomegaly
    - Pulmonary vascular markings
    - Pulmonary vascular disease
- ECG
    - Routine (left ventricular hypertrophy [LVH], right ventricular hypertrophy [RVH], cardiovascular hypertrophy [CVH])
    - Superior QRS-axis
    - Right bundle branch block
    - Conductance disturbances
- ECHO/TOE
    - Most useful investigation for
        - Left AV valve morphology and function
        - Ventricular function
        - Residual lesions (shunt, subaortic stenosis)
- Catheter
    - Rarely required unless reoperation considered
- MRI
    - Rarely indicated
- Holter monitor
    - Only in symptomatic patients
- Exercise testing
    - Rarely indicated
- Additional investigations
    - Significant left AV valve dysfunction
    - Significant residual shunt
    - Subaortic stenosis

Indications for reintervention

- Significant left AV-valve dysfunction
- Significant residual shunt
- Subaortic stenosis
- Progressive/symptomatic AV-Block

Interventional options

- Reoperation may require valve replacement

Posttreatment outcome

- Excellent long-term results unless
    - Actuarial survival after 20 years >80%
    - Left AV valve regurgitation (stenosis)
    - Pulmonary vascular disease
    - Late subaortic stenosis

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception

- Pregnancy contraindicated in PVD (Eisenmenger)
- Anticoagulation management in patients with prosthetic valves
- Avoid oestrogen-containing pill in pulmonary hypertension

Recurrence/genetics/syndromes

- Above average recurrence risk Down´s syndrome in >50% of complete AVSD
- Approximately 10 to 14% congenital cardiac defects in mothers with AVSD

Physical activity/sports

- No restrictions if good repair and no significant arrhythmias

Insurance

- Category 2 if well repaired

Follow-up interval

- 1 to 2 yearly intervals with ECG and ECHO in stable cases

Follow-up care

- Level 2 unless significant haemodynamic problems

Unresolved issues

- Only limited data regarding long-term prognosis

Postoperative partial atrioventricular septal defect (p-AVSD)

Survival/adult life

- Similar to secundum atrial septal defect unless significant to left AV-valve regurgitation
- Unoperated p-AVSD have reduced life expectancy
- Pulmonary veno-occlusive disease (PVOD) may develop late
- Status after repair depends on left AV valve function

Hemodynamic issues

- Before repair
    - Size of shunt
    - Degree of AV valve regurgitation
- After repair
    - Residual shunt and left AV valve regurgitation
    - Subaortic stenosis

Arrhythmia/pacing

- Atrial arrhythmias rare unless left AV-valve regurgitation
- Complete heart block very rare
- Pacing rarely required unless sick sinus syndrome

Investigations

- ECG
    - Superior-QRS axis
    - Right bundle branch block
    - Rhythm follow-up
- Chest x-ray
    - Routine
    - Cardiomegaly
- ECHO/TOE
    - Most useful investigation both for and after operation
    - Left AV valve function
    - Ventricular function
    - Residual lesion
- Catheter
    - Rarely required unless reoperation considered
- MRI
    - Rarely indicated
- Holter monitor
    - Rarely indicated
- Exercise testing
    - Rarely indicated
- Additional investigations
    - None

Indications for intervention

- All cases to be considered for intervention unless pulmonary vascular disease
- Reoperation for significant left AV-valve regurgitation
- Residual shunt or subaortic stenosis
- Progressive/symptomatic arrhythmias

Interventional options

- Surgery with valve repair or replacement
- Closure of re-/residual ASD
- Pacemaker in progressive/symptomatic complete block

Outcome

- Excellent long term provided left AV-valve repair satisfactory

Endocarditis

- Prophylaxis indicated if left AV-valve regurgitation is present

Pregnancy/contraception

- Well tolerated in repaired cases
- Contraindicated in rare cases with PVOD (Eisenmenger)
- Anticoagulation management in patients with prosthetic valves
- Avoid oestrogen-containing pill in pulmonary hypertension

Recurrence/genetics/syndromes

- None

Physical activity/sports

- No restrictions if good repair and no significant arrhythmias

Insurance

- Category 2

Follow-up interval

- 2 yearly intervals with ECG and ECHO in stable cases

Follow-up care

- Unoperated level 1; postoperative level 2

Unresolved issues

- Long-term function of a nonreconstructed AV-valve is uncertain

Pulmonary stenosis (PS)

Survival/adult life

- Excellent if relieved effectively
- Poor if severe valve PS untreated

Haemodynamic issues

- PS severity
- Pulmonary regurgitation (PR) severity
- Leaflet dysplasia
- Right ventricular function

Arrhythmia/pacing

- Atrial arrhythmias in RV failure and tricuspid regurgitation
- Pacing not indicated

Investigations

- Chest x-ray
    - Baseline; otherwise little value unless RV failure
- ECG
    - Rhythm RV
    - Hypertrophy
- ECHO/TOE
    - Investigation of choice for right ventricular outflow tract (RVOT) gradient, pulmonary regurgitation RV size/function, tricuspid regurgitation
- Catheter
    - Rarely needed except for balloon dilatation
- MRI
    - Rarely needed
    - Assess RV size/function and right atrial (RA) dilation in severe pulmonary regurgitation
- Holter monitor
    - Not routinely indicated
- Exercise
    - Not routinely indicated
- Additional investigations
    - None

Indications for intervention

- Valve gradient >30 mmHg at rest or for symptoms

Interventional options

- Balloon valvuloplasty almost always
- Surgery if valve calcified/dysplasty

Posttreatment outcome

- Excellent long-term results unless early failure

- Significant pulmonary regurgitation uncommon

Endocarditis

- Low risk. Prophylaxis may not be required in mild cases

Pregnancy/contraception/fetal

- Routine pregnancy unless moderate to severe PS or right to left shunt through ASD or patent foramen ovale (PFO)

Recurrence/genetics

- 4% approximately

Syndromes

- Noonan
- Congenital rubella
- Williams
- Alagille

Sport/physical activity

- Unrestricted unless severe

Insurance

- Category 1 after successful treatment or mild PS

Follow-up interval

- Can discharge if mild with ECHO. Every 1 to 3 years if more than mild, PR, or desaturation.

Follow-up care

- Mild PS: 3; excellent early result: 2; residual gradient or significant PR: 2

Unresolved issues

- None

Tetralogy of Fallot – postoperative

Introduction and background

- Common lesion. Most Fallot patients are now adults.

Survival/adult life

- Survival rate after surgery excellent (normal in selected groups)
- Occasionally unoperated patients survive into adulthood.

Haemodynamic issues

- Pulmonary regurgitation/PS and RV function
- Tricuspid regurgitation
- Aortic regurgitation
- Residual lesions

Arrhythmia/pacing

- Late complete heart block rare
- Ventricular premature beats common in asymptomatic patients
- Symptomatic ventricular tachyarrhythmias (VT) rare
- Atrial arrhythmias common and relate to poor haemodynamics
- Small incidence of late sudden death

Investigations

- Chest x-ray
    - Baseline and occasionally follow-up
    - Cardiomegaly
    - RV outflow
- ECG
    - Routine
    - Rhythm
    - access/QRS width (usually complete right bundle branch block)
- ECHO/TOE
    - Regularly for PR/RVOT/RV size function/tricuspid regurgitation
    - Aortic regurgitation/LV function
- Catheter
    - Preoperative for residual lesions, coronary anatomy intervention for dilatation/stent of pulmonary arteries
    - Possibly in future for implantable pulmonary valve
- MRI
    - May become investigation of choice for RV size function and pulmonary regurgitation
- Holter monitor
    - For symptoms and in poor haemodynamics
- Exercise
    - Exercise capacity, arrhythmias
- Additional investigations
    - Electrophysiological study for syncope, sustained arrhythmia (atrial or ventricular), radiofrequency ablation (RFA)

Indications for intervention

- Significant RVOT or PA branch stenosis

- Aortic regurgitation
- Residual VSD, significant pulmonary regurgitation (with symptoms and RV dilatation)

Interventional options

- Surgery, surgery with ablation, balloon dilatation/stenting, RFA catheter intervention for pulmonary valve insertion

Posttreatment outcome

- Most patients well
- RV function may not normalize after pulmonary valve replacement
- Arrhythmia may persist
- Risk of sudden death

Endocarditis

- Prophylaxis in all

Pregnancy/contraception/fetal

- No contraindication to pregnancy in well repaired patients
- Monitor ventricular function and arrhythmia
- No additional fetal risk

Recurrence/genetics

- 1.5% for father, 2.5 to 4% for mother with Tetralogy of Fallot
- 16% of Fallot patients have deletion of chromosome 22q11; recurrence risk 50%.

Syndromes

- 22q11

Sport/physical activity

- No contraindication to sport unless documented arrhythmia
- Significant ventricular dysfunction

Insurance

- Category 2

Follow-up interval

- One/two yearly with ECG, ECHO $\pm$ Holter, exercise test

Follow-up care

- 1 if documented residual abnormalities/arrhythmia, 2 otherwise

Unresolved issues

- Risk stratification for sudden death
- Indication for implantable defibrillator
- Timing of reoperation for pulmonary regurgitation

Conduits

Introduction and background

- Conduits used in repair of complex congenital heart disease
- Usually RV-PA (e.g. PA/VSD, Truncus, Tetralogy of Fallot, transposition of great artery/VSD/PS)

Survival/adult life

- All conduits in children deteriorate and require replacement (usually <10 years)
- Longevity of replacement unclear

Haemodynamic issues

- Stenosis of valve, subvalve, or anastomosis to PA
- Pulmonary regurgitation with RV volume overload
- LV-aortic pathway in complex repairs

Arrhythmia/pacing

- Ventricular arrhythmias, surgical heart-block

Investigations

- Chest x-ray
    - Baseline and follow-up
    - Conduit calcification
    - Cardiomegaly
- ECG
    - Routine
    - Rhythm
- ECHO
    - Investigation of choice for follow-up of RV
    - Pressure gradient across conduit and pacing impulse
    - LV aortic pathway
    - ECHO may underestimate gradient.
- TOE
    - Not routine
- Catheterization
    - Evaluation for surgery
    - Balloon dilatation or stenting

- MRI
    - Very useful for investigation of conduit function
    - May become investigation of choice
- Holter monitor
    - Only if arrhythmia suspected
- Exercise function
    - Not routine
    - Useful for objective evaluation of exercise tolerance

Indications for intervention

- Significant symptoms or conduit obstruction

Interventional options

- Usually surgical replacement of conduits
- Occasionally balloon dilatation or stenting

Posttreatment outcome

- Fate of replaced conduit uncertain
- Need long-term follow-up

Endocarditis

- Prophylaxis in all

Pregnancy/contraception

- Pregnancy tolerated if haemodynamics stable
- No contraception issues

Recurrence/genetics

- Usual recurrence rate for congenital heart disease
- Higher if 22q11 deletion

Syndromes

- 22q11 deletion

Sport/physical activity

- Avoid contact sports
- Otherwise no restrictions if haemodynamics good

Insurance

- Level 2

Follow-up interval

- Yearly with ECHO, ETT for ventricular function, arrhythmia surveillance (ECG, Holter if symptoms)
- Significant conduit dysfunction may be present in mildly symptomatic patients

Follow-up care

- Category 1

Unresolved issues

- Type of conduit (homograft versus xenograft)
- Role of balloon dilatation stenting

Aortic valve stenosis (unoperated)

Introduction and background

- Common, especially bicuspid aortic valve (1–2% of population)
- May occur with other lesions

Survival/adult life

- Normal if mild obstruction

Hemodynamic issues

- Degree of stenosis may progress
- Associated aortic regurgitation
- LV hypertrophy and function

Arrhythmia/pacing

- Ventricular tachyarrhythmias (VT) and ventricular fibrillation (VF) may occur during exertion with severe obstruction

Investigations

- ECG
    - LVH and repolarization changes
- Chest x-ray
    - Baseline
    - Calcification
- ECHO
    - Investigation of choice
    - LV mass/function
    - Aortic valve/size/morphology/area
    - LV to aortic gradient
    - Aortic regurgitation

- TOE
    - Rarely of value except in endocarditis
- MRI
    - Rarely of value
- Catheter
    - Not for diagnosis
    - For coronary angiography and balloon dilatation
- Exercise testing
    - For repolarization changes and symptoms
    - Surgical decision making

Indications for intervention

- Symptoms: severe LV pressure overload
- Severe aortic stenosis

Interventional options

- Balloon valvuloplasty if valve uncalcified
- Rarely good option in adult
- Mechanical valve replacement, homograft, or Ross procedure depending on patient's age, sex, preferences, and local expertise

Outcome

- Recurrence common late after valvotomy
- Very good in uncomplicated cases of valve replacement.

Endocarditis

- Prophylaxis indicated in all

Pregnancy/contraception

- Low risk in asymptomatic patents even with moderate obstruction
- High risk in patients with severe obstruction
- Transcatheter intervention may be indicated in unplanned pregnancy

Recurrence/genetics/syndromes

- Bicuspid valve may be familial
- Association with coarctation
- Recurrence rate may be higher in syndromes.

Physical activity/sports

- No competitive sports if obstruction is moderate or severe

Insurance

- Category 2

Follow-up interval

- Depends on severity and progression rate ECG/ECHO ± exercise test

Follow-up care

- Mild 3-moderate/severe 1

Unresolved issues

- Late outcome after the Ross operation

Postoperative valvar aortic stenosis

Introduction and background

- Common lesion
- Most interventions in children are balloon dilation or open aortic valvotomy; aortic valve replacement, mechanical or biological prostheses, or Ross procedure may have been performed.

Survival — adult life

- Excellent

Hemodynamic issues

- Obstruction
- Regurgitation
- LV function
- Pulmonary homograft (Ross)

Arrhythmia/pacing

- Arrhythmia rare
- More common in left ventricular hypertrophy (LVH)
- May cause sudden death

Investigations

- ECG
    - Routine LVH
    - Conduction disturbances
    - Repolarization changes
- Chest x-ray
    - Cardiomegaly
- ECHO
    - See "Aortic valve stenosis (unoperated)"

    - Prosthesis function and paravalvular leak
- TOE
    - Useful in assessment of paravalvular leaks and suspected endocarditis
- MRI
    - Rarely indicated
- Catheter
    - Rarely indicated (see "Aortic valve stenosis [unoperated]")
- Exercise testing
    - Surgical decision making for timing of reintervention

Indications for reintervention

- Recurrent obstruction (native valve or prosthesis)
- Regurgitation
- Occasionally haemolysis

Interventional options

- Mechanical valve, homograft, or Ross operation
- Prosthesis may be preferred by elderly.
- Homograft may be preferred in endocarditis.

Outcome

- Very good but anticoagulant problems with mechanical valve and late failure

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception

- Anticoagulants may cause embryopathy.

Recurrence/genetics/syndromes

- See "Aortic valve stenosis (unoperated)"

Physical activity/sports

- High-level activity possible in uncomplicated cases with good LV function
- Contact contraindicated in patients on anticoagulants

Insurance

- Category 2

Follow-up interval

- Yearly

Follow-up care

- Ross 1; otherwise 2

Unresolved issues

- Long-term outcome of Ross procedure
- Best anticoagulation protocol in pregnancy

Subaortic stenosis unoperated

Introduction and background

- Uncommon form of obstruction
- May be discrete or extend to adjacent structures
- Often progressive.

Survival/adult life

- Normal if obstruction not severe

Hemodynamic issues

- Progression very common
- May cause aortic regurgitation
- Associated lesions common (e.g., VSD)

Arrhythmia/pacing

- See "Aortic valve stenosis"

Investigations

- Chest x-ray
    - Baseline
- ECG
    - Routine LVH and repolarization changes
- ECHO
    - Investigation of choice
    - Visualise obstruction
    - Gradient across LV outflow tract
    - LV mass/function
    - Aortic regurgitation
- TOE
    - May be useful to define anatomy
- MRI
    - Rarely indicated
- Catheter
    - Rarely indicated (see "Aortic valve stenosis")
- Exercise test
    - For repolarization changes and symptoms

Indications for intervention

- Progressive obstruction
- Lower threshold and aortic valve stenosis
- Aortic regurgitation

Interventional options

- Surgical resection

Outcome

- Recurrence possible

Endocarditis

- Prophylaxis in all

Pregnancy/contraception

- Low risk if no severe obstruction

Recurrence/genetics/syndromes

- May occur left heart abnormalities (e.g., coarctation, Shone´s syndrome)
- Familial cases described

Physical activity/sports

- No restriction if mild obstruction or after resection

Insurance

- Category 2

Follow-up interval

- Depends on severity and progression rate; usually 1 to 2 yearly

Follow-up care

- Level 1

Unresolved issues

- Recurrence rate after resection
- Optimal timing of surgery

Unoperated coarctation

Introduction and background

- May present in infancy or later in adolescence

Survival/adult life

- Rarely undiagnosed in childhood, but long-term survival is possible

Haemodynamic issues

- Hypertension
- Premature atherosclerosis
- LV hypertrophy/failure
- Aortic dissection
- Associated aortic/mitral valve (MV) lesions

Arrhythmia/pacing

- Rare problems

Investigations

- ECG
    - LVH repolarization changes
- Chest x-ray
    - Cardiomegaly
    - Ascending aorta dilation
    - Rib notching
- ECHO
    - Assessment of arch anatomy/gradient
    - Associated lesions LVH and function
- TOE
    - Rarely provides additional information
- MRI
    - Investigation of choice
- Holter monitor
    - Not indicated unless for ambulatory blood pressure
- Exercise test
    - Hypertension on exercise
    - Arm/leg gradient
    - Inducible repolarization abnormalities
- Catheterization
    - If MRI unavailable for arch anatomy
    - For coronary angiography when indicated for intervention
- Additional
    - Screen for intracerebral vascular anomalies

Indications for intervention

- Resting or exercise induced hypertension
- Resting gradient >30 mmHg

Interventional options

- Balloon/stenting
- Surgical repair

Posttreatment outcome

- Residual hypertension common despite adequate relief of obstruction
- Accelerated atherosclerosis
- Reduced life expectancy

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception/recurrence/fetal

- Repair prior to pregnancy if possible
- Transcatheter intervention may be indicated in unplanned pregnancy (worsening blood pressure [BP], LV failure)
- Avoid oestrogen containing pill
- Growth retardation common
- Spontaneous foetal loss increased

Recurrence/genetics

- Recurrence may be familial
- 22q11 deletion in complex forms

Syndromes

- Turner´s (present in approx 30%)
- Williams´ (present in approx 10%)
- Shone´s (associated LV inflow/outflow abnormalities)

Sport/physical activity

- Should be restricted prior to repair

Insurance

- Category 3 for significant unoperated coarctation

Follow-up interval

- Most patients referred for intervention on diagnosis
- 1 yearly of mild cases with BP at rest and exercise/ECHO/Doppler/MRI

Follow-up care

- Level 1

Unresolved issues

- Influence of age at operation on long-term outcome
- Influence of drugs on vascular phenotype in successful cases
- Role of intervention for mild gradients
- Role of stenting as adjunct to balloon

Operated coarctation

Survival/adult life

- Long-term survival still reduced despite adequate early repair

Haemodynamic issues

- Persistent and late developing hypertension at rest and exercise
- Aortic valve dysfunction
- Rare dissection

Arrhythmia/pacing

- Not an issue

Investigations

- ECG
    - LVH ± repolarization changes
- Chest x-ray
    - Cardiomegaly
    - Ascending aorta dilation
    - Rib notching
- ECHO
    - Assessment of arch anatomy/gradient
    - Associated lesions LVH and function
- TOE
    - Rarely provides additional information
- MRI
    - Investigation of choice
- Holter monitor
    - Not indicated unless for ambulatory blood pressure
- Exercise test
    - Hypertension on exercise
    - Arm/leg gradient
    - Inducible repolarization abnormalities
- Catheterization
    - If MRI unavailable for arch anatomy
    - For coronary angiography when indicated for intervention
- Additional
    - Screen for intracerebral vascular anomalies advocated by some

Indications for intervention

- Significant recoarctation (gradient >30 mmHg at rest)
- Aortic aneurysm

Interventional options

- Balloon/stenting for anatomically suitable recoarctation
- Surgery for complex situations ± aneurysms

Posttreatment outcome

- Excellent but late hypertension and premature atherosclerosis/cerebrovascular accident/myocardial infarction/heart failure

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception/recurrence/fetal

- Relieve residual coarctation prior to pregnancy or during unplanned pregnancy
- Monitor closely for hypertension
- Avoid oestrogen-containing pill if rest or exercise hypertension

Recurrence/genetics

None stated

Syndromes

None stated

Sport/physical activity

- No restrictions if adequate relief of obstruction/no residual hypertension

Insurance

- Category 2

Follow-up interval

- Yearly with same investigations as for unoperated coarctation

Follow-up care

- Level 2

Unresolved issues

- Influence of age at repair, type of repair of intervention on late hypertension
- Late outcome of balloon/stenting
- Pathophysiology of late hypertension

Patent arterial duct

Survival/adult life

- Normal life expectancy in closed patent ductus arteriosus (PDA)
- Rare PVD for large PDA

Haemodynamic issues

- Usually none — LV dilatation/pulmonary hypertension in significant PDA

Arrhythmia/pacing

- None

Investigations

- Chest x-ray
    - Baseline
    - Cardiomegaly
    - Ductal calcification
- ECG
    - Usually normal
    - LVH with large PDA
- ECHO/TOE
    - Usually diagnostic
    - TOE rarely indicated
- Catheter
    - For closure coronary angiography in older patients
- MRI
    - Not indicated
- Holter monitor
    - Not indicated
- Exercise
    - Not indicated
- Additional investigations
    - None

Indications for intervention

- Controversial for silent of very small PDA
- Continuous murmur
- LV dilatation

Interventional options

- Catheter closure intervention of choice
- Several device options
- Surgery for rare cases

Posttreatment outcome

- Excellent
- Residual shunt in up to 10%

Endocarditis

- Not required after complete closure
- Prophylaxis indicated otherwise

Pregnancy/contraception/fetal

- No problems unless pulmonary vascular disease

Recurrence/genetics

- None

Syndromes

- Congenital rubella

Sport/physical activity

- No restrictions unless PVD

Insurance

- Category 1 for small PDA or after closure

Follow-up interval

- Discharge 1 year after closure

Follow-up care

- Level 3 unless PVD (1)

Unresolved issues

- Indication of closure for small PDA

Ebstein´s anomaly

Introduction and background

- Wide spectrum of pathologic anatomy which determines onset of severity of symptoms

Survival/adult life

- Extremely variable natural history
- Infant survivors usually reach

Haemodynamic issues

- Cyanosis at rest and/or exercise (right-left shunt at atrial level), reduced exercise capacity
- Congestive heart failure (tricuspid stenosis/regurgitation/small RV)
- Associated lesions
- LV abnormalities

Arrhythmia/pacing

- Atrial arrhythmias are common.
- Increase with age
- Related to pre-exultation and atrial dilatation
- Risk of sudden death

Investigations

- Chest x-ray
    - Marked cardiomegaly
    - Right atrial enlargement
- ECG
    - Baseline (characteristic pattern)
    - Follow-up for rhythm
- ECHO/TOE
    - Severity of tricuspid valve displacement dysplasia and regurgitation
    - RV size
    - Associated lesions
    - LV function
- Catheter
    - Rarely required unless for coronary angiography in older patients or at electrophysiologic studies (EPS)
- MRI
    - Rarely required
- Holter monitor
    - Useful for arrhythmia monitoring
- Exercise
    - Baseline and follow-up
    - Cyanosis
    - Exercise tolerance
    - Arrhythmia
- Additional investigations
    - Electrophysiologic studies (EPS) for arrhythmia diagnosis and RFA

Indications for intervention

- Decrease in exercise tolerance
- Heart failure
- Increase in cyanosis
- Arrhythmia

Interventional options

- Surgery for tricuspid valve repair or replacement
- RFA for arrhythmias/preexultation

Posttreatment outcome

- Symptomatic improvement usual
- Tricuspid valve replacement: reoperation, thrombotic complications
- Ongoing arrhythmia problems frequent
- Risk of sudden death remains
- Anticoagulants for atrial arrhythmia and prosthetic tricuspid valve

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception/fetal

- Well tolerated unless cyanosis or heart failure
- Foetus at risk in cyanosed mother

Recurrence/genetics

- 6% in affected mother; 1% in affected father. Familiar occurrence documented.

Syndromes

- Rare

Sport/physical activity

- Recreational sport in asymptomatic patient

Insurance

- Unoperated asymptomatic or well postoperative category 2

Follow-up interval

- Depends on clinical status
- Annual follow-up with ECHO/Holter exercise test

Follow-up care

- Level 1 (operated and unoperated)

Unresolved issues

- Recurrence of arrhythmias
- Long-term fate of repairs

Fontan

Introduction and background

- Palliative procedure for single ventricle physiology in which all systemic venous return directed to the lungs; multiple modifications

Survival/adult life

- Improved survival with strict selection criteria
- Late failure even in best cases

Haemodynamic issues

- Function of systemic ventricle (preload deproved)
- Pulmonary vascular resistance
- Obstruction in Fontan connection
- Atrial enlargement
- Pulmonary venous obstruction
- AV valve regurgitation
- Chronic venous hypertension
- Desaturation/paradoxical embolus in fenestrated Fontan
- Pulmonary arteriovenous malformations in some

Arrhythmia/pacing

- Atrial arrhythmias common
- Increase with follow-up
- Sinus node dysfunction
- Pacing: ventricular pacing requires epicardial system

Investigations

- Chest x-ray
    - Baseline and follow-up
    - Cardiomegaly
    - Pulmonary vascular markings
- ECG
    - Rhythm
- ECHO/TOE
    - Most useful investigation for
        - ventricular function

        - AV valve regurgitation
        - Residual shunts
        - Obstruction of Fontan connections
        - Thrombus in atrium
    - Routine TOE (2 yearly may be indicated or if arrhythmia present)
- Catheter
    - For haemodynamic assessment and angiography in clinical deterioration
- MRI
    - Obstruction of Fontan connection
    - Occasionally useful for RA size and anastamieos
- Holter monitor
    - Routine and for symptomatic arrhythmia
- Exercise testing
    - Reaction activities only
- Additional investigations:
    - Blood/stool for protein-losing enteropathy (PLE)

Indications for intervention

- Cyanosis
- Obstruction to Fontan connection
- Systemic AV valve regurgitation
- Ventricular failure
- Arrhythmia
- Pulmonary venous obstruction

Interventional options

- Consider conversion to total cavopulmonary connection (TCPC) or transplant in failing Fontan
- Closure of fenestration
- AV malformations
- RFA
- Supraventricular arrhythmia
- AV sequential pacing

Posttreatment outcome

- Variable success with catheter ablation of atrial arrhythmias
- PLE has <50% 5-year survival
- Fontan conversion results unclear; atrial arrhythmias common.

Endocarditis

- Prophylaxis in all

Pregnancy/contraception/fetal

- Pregnancy possible with perfectly selected patients and proper care
- High maternal risk in "failing Fontan"

- Higher miscarriage rate
- Foetal risk of congenital heart disease (CHD) may be higher
- Avoid oestrogen pill if ejection fraction <40%, residual shunt, or spontaneous contrast in RA
- Angiotensin-converting enzyme (ACE) inhibitors should be withdrawn if on anticoagulants: need meticulous management

Recurrence/genetics

- None

Syndromes

- None

Sport/physical activity

- Recreational sports only

Insurance

- Category 3

Follow-up interval

- At least yearly review with ECHO, ECG, Holter, exercise testing, blood testing

Follow-up care

- Level 1

Unresolved issues

- Indications for and results of Fontan conversion
- Outcome of TCPC in modern era
- Role of anticoagulation
- Medical therapy for failing systemic ventricle
- Role of ACE inhibitors

Marfan´s syndrome

Introduction and background

- Abnormal fibrillin gene on chromosome 15q
- Autosomal dominant inheritance
- Cardiac defect largely determined outcome

Survival/adult life

- Death from cardiac problems

- Life expectancy reduced but improved by good cardiac follow-up and surgery

Haemodynamic issues

- Acute aortic dissection; risk higher if the aortic sinuses >55 mm
- Aortic regurgitation
- MV prolapse/regurgitation

Arrhythmia/pacing

- Atrial and ventricular arrhythmia in MV prolapse/regurgitation

Investigations

- Chest x-ray
    - Not helpful for follow-up of aorta
- ECG
    - Rarely useful
- ECHO/TOE
    - Most valuable investigation for serial follow-up of aortic root dimensions and valve function (aortic and mitral)
- Catheter
    - Rarely indicated
- MRI
    - Excellent investigation for aortic arch and descending aorta
    - Compliments echocardiography
- Holter
    - Not routine
- Exercise testing
    - Not routine
- Additional investigations
    - Noncardiac assessment (e.g., ophthalmic, orthopaedic)

Indications for intervention

- Beta blockers for aortic dilatation
- Surgery if aortic diameter >55 mm or rapid increase
- Significant aortic regurgitation
- Significant mitral regurgitation

Interventional options

- Urgent surgery for dissection
- Aortic root and valve replacement
- Valve sparing operation may be indicated

Posttreatment outcome

- Surgery improves life expectancy but other dissections still possible
- Beta blockers delay/prevent progression

Endocarditis

- Prophylaxis in valve regurgitation and after aortic surgery

Pregnancy/contraception/fetal

- Pregnancy contraindicated if aorta is >45 mm
- Pregnant women should be on beta blockers.
- Caesarean section to be discussed if aorta is dilated

Recurrence/genetics/syndrome

- Approximately 50% (autosomal dominant)

Sport/physical activity

- Strenuous exercise contraindicated
- High altitude and diving contraindicated (spontaneous pneumothorax)

Insurance

- Category 3

Follow-up interval

- Annual follow up for aortic dilatation
- More frequent evaluation if aortic diameter increasing

Follow-up care

- Level 1

Unresolved issues

- Role of early beta blockade
- Long-term results of surgery including valve sparing

Postoperative transposition (Mustard/Senning)

Introduction and background

- Common lesion: most Mustard/Sennings patients now adults; operation replaced by arterial switch mid 1980´s

Survival/adult life

- Low early mortality
- Significant late morbidity/mortality from arrhythmia/baffle obstruction/RV failure with risk of sudden death

Haemodynamic issues

- Intraatrial baffle obstruction (systemic and pulmonary venous) more common in Mustard than Senning
- Tricuspid regurgitation/RV failure relatively rare but important to detect early

Arrhythmia/pacing

- Progressive loss of sinus rhythm on Holter with follow-up
- Slow junctional rhythm may rarely require pacing.
- Tachyarrhythmias (predominantly atrial flutter) may be related to high incidence of late sudden death.
- Pacing may be required if antiarrhythmic drugs

Investigations

- ECG
    - Right ventricular hypertrophy with basic rhythm (often junctional)
- Chest x-ray
    - Useful for cardiomegaly
    - Pulmonary venous obstruction
- ECHO/TOE
    - TTE for ventricular function/tricuspid regurgitation
    - TOE essential if questions remain regarding baffle function
- MRI
    - Rarely required if TOE available
- Holter monitor
    - Occult arrhythmia
    - Not predictive of septal defect (SD)
- Exercise test
    - Exercise tolerance
    - Evaluation of arrhythmia
- Catheterization
    - For intervention and assessment of new onset symptoms
- Additional
    - Electrophysiologic study/RFA for refractory atrial arrhythmias

Indications for intervention

- Baffle obstruction
- Baffle leaks
- Tricuspid valve dysfunction
- RV failure

Interventional options

- Balloon/stenting for pathway obstruction
- Transcatheter closure for baffle leaks
- Tricuspid valve/replacement
- Conversion to arterial switch (pulmonary artery banding)
- Transplantation

Posttreatment outcome

- Risk of sudden death despite lack of symptoms or overt haemodynamic disturbance

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception/recurrence/fetal

- Pregnancy not contraindicated in most cases
- Monitor RV function throughout
- No contraceptive issues
- Long-term consequences on RV function not known

Recurrence/genetics

- Familial recurrence of transposition of great artery rare

Syndromes

- None

Sport/physical activity

- Generally normal activities
- Maximal exercise tolerance likely to be diminished

Insurance

- Category 3

Follow-up interval

- Yearly

Follow-up care

- Level 1

Unresolved issues

- Risk stratification for sudden death
- Fate of systemic RV/tricuspid valve
- Indication/conversion/transplant strategies

Congenitally corrected transposition

Introduction and background

- Rare lesion
- Usually associated with other abnormalities
- May occur with dextrocardia

Survival/adult life

- Common to survive to adult life
- Associated lesions common (VSD, PS, left AV valve regurgitation) determine outcome

Haemodynamic issues

- Cyanosis with VSD and PS
- PVD if VSD and no PS
- Systemic ventricular failure with systemic AV valve regurgitation
- Referral before systemic ventricular dysfunction

Arrhythmia/pacing

- Spontaneous complete heart block (2% per year) and postsurgical heart block
- Endocardial pacing in the morphologic LV
- Atrial arrhythmias common
- Ventricular arrhythmias with systemic ventricular dysfunction
- Epicardial pacing if potential for paradoxical embolus

Investigations

- Chest x-ray
    - Baseline
    - Follow-up for associated lesions
    - Cardiomegaly
- ECG
    - Rhythm
- ECHO/TOE
    - Size and function of systemic ventricle
    - Morphology of left AV valve
    - Associated lesions
- Catheter
    - For pulmonary haemodynamics and anatomy of associated lesions
- MRI
    - Rarely required
- Holter monitor
    - For occult arrhythmia detection
- Exercise function
    - Helpful for timing of surgery
    - Oximetry
    - Exercise tolerance
- Additional investigations
    - Occasionally multi-gated acquisition scan (MUGA) for ventricular function

Indications for intervention

- +>Moderate systemic AV valve regurgitation
- Significant associated lesions
- Pacemaker for complete AV block with symptoms, profound bradycardia, or chronotropic incompetence

Interventional options

- Valve replacement
- Pulmonary artery banding
- "Double switch" (controversial in adults)

Posttreatment outcome

- Good if left AV valve replacement before systemic ventricular function deteriorates
- Atrial arrhythmias common

Endocarditis

- Prophylaxis in all cases

Pregnancy/contraception/fetal

- Pregnancy not contraindicated if asymptomatic
- Monitor ventricular function and rhythm
- Long-term consequences on systemic ventricular function unknown
- Avoid oestrogen-containing contraceptive pill if cyanosed/pulmonary hypertension

Recurrence/genetics

- 4%

Syndromes

- None

Sport/physical activity

- No restriction on recreational activities

Insurance

- Category 3 in most cases

Follow-up interval

- Yearly, with ECHO, exercise test ± Holter

Follow-up care

- Level 1 (pre and postoperative)

Unresolved issues

- Classical repair of VSD and PS versus "double switch."

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is not specifically stated for each recommendation. In the section of the guideline regarding specific lesions (section 7), many recommendations are based on clinical experiences rather than evidence from randomized clinical trials.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Appropriate and effective management of congenital heart disease in adults
- Reduction in avoidable medical problems and adult deaths from congenital heart disease
- The establishment of specialized centres to manage the complex grown-up heart disease population

POTENTIAL HARMS

- Low-dose oestrogen combined oral contraceptive pills are very efficacious, but their thrombogenic properties may make them hazardous in certain situations, such as after the Fontan operation or in patients with atrial fibrillation/flutter.
- The oestrogen-containing contraceptive pill should not be used in patients with pulmonary or systemic hypertension. Medroxyprogesterone injection (Depo-Provera®), subcutaneous deposition of levonorgestrel (Norplant®), or progesterone only pills are effective, but may cause fluid retention and should not be used in patients with heart failure.

CONTRAINDICATIONS

CONTRAINDICATIONS

- Eisenmenger patients are at particular risk from pregnancy, dehydration, cardiac and non-cardiac surgery, general anaesthesia, anaemia, chest

infections, altitude, intravenous lines and vasodilating drugs. There is approximately a 50% maternal fatality in conjunction with pregnancy, and death often occurs after delivery. Pregnancy is therefore contraindicated, even in those with "moderate" pulmonary hypertension. Combined oral contraceptives are contraindicated in patients at risk of paradoxical embolism, unless they are also receiving anticoagulants

- Marfan syndrome: strenuous exercise contraindicated; high altitude and diving contraindicated (spontaneous pneumothorax).

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

Strategies for delivery of patient care and training of practitioners for grown-up congenital heart disease are provided in the original guideline document.

IMPLEMENTATION TOOLS

Personal Digital Assistant (PDA) Downloads
Pocket Guide/Reference Cards
Resources
Slide Presentation

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Safety

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

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GUIDELINE COMMITTEE

Task Force on the Management of Grown Up Congenital Heart Disease of the European Society of Cardiology

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Task Force Members: John Deanfield, Chairperson; Erik Thaulow; Carol Warnes; Gary Webb; Frantizek Kolbel; Andreas Hoffman; Keld Sorenson; Harald Kaemmerer; Ulf Thilen; Margreet Bink-Boelkens; Laurence Iserin; Luciano Daliento; Eric Silove; Andrew Redington; Pascal Vouhe

European Society of Cardiology Committee for Practice Guidelines (CPG) Members: Silvia Priori, Chairperson; Maria Angeles Alonso; Jean-Jacques Blanc; Andrzej Budaj; Martin Cowie; Jaap Deckers; Enrique Fernandez Burgos; John Lekakis; Bertil Lindahl; Gianfranco Mazzotta; Joao Morais; Ali Oto; Otto Smiseth; Hans Joachim Trappe

Document Reviewers: Jaap Deckers, CPG Review Coordinator; Werner Klein, Former CPG Chairperson; Maria Angeles Alonso; Carina Blomstrom-Lundqvist; Guy de Backer; Jaromir Hradec; Gianfranco Mazzotta; Alexander Parkhomenko; Patrizia Presbitero; Adam Torbicki

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ENDORSER(S)

Armenian Society of Cardiology - Medical Specialty Society
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Finnish Cardiac Society - Medical Specialty Society
French Society of Cardiology - Medical Specialty Society
German Society of Cardiology - Medical Specialty Society
Hellenic Cardiological Society - Medical Specialty Society
Israel Heart Society - Medical Specialty Society
Italian Federation of Cardiology - Medical Specialty Society
Latvian Society of Cardiology - Medical Specialty Society
Lithuanian Society of Cardiology - Medical Specialty Society
Netherlands Society of Cardiology - Medical Specialty Society
Norwegian Society of Cardiology - Medical Specialty Society
Portuguese Society of Cardiology - Medical Specialty Society
Romanian Society of Cardiology - Medical Specialty Society
San Marino Society of Cardiology - Medical Specialty Society
Society of Cardiology of the Russian Federation - Medical Specialty Society
Spanish Society of Cardiology - Medical Specialty Society
Swiss Society of Cardiology - Medical Specialty Society
Turkish Society of Cardiology - Medical Specialty Society

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available from the European Society of Cardiology (ESC) Web site.

Print copies: Available from Elsevier Publishers Ltd., 32 Jamestown Road, London, NW1 7BY, United Kingdom. Tel: +44.207.424.4422; Fax: +44 207 424 4433; E-mail: gr.davies@elsevier.com

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- Management of grown-up congenital heart disease. Pocket guidelines. Available from the ESC Web site. Also available for PDA download from the ESC Web site.
- Management of grown-up congenital heart disease. Slide set. Available from the ESC Web site.
- Adult congenital heart disease survival simulator. Available from the ESC Web site.
- Recommendations for guidelines production. A document for Task Force Members responsible for the production and updating of ESC guidelines. 2006 Jun 28. 21 p. Available in Portable Document Format (PDF) from the ESC Web site.

Print copies: Available from Elsevier Publishers Ltd., 32 Jamestown Road, London, NW1 7BY, United Kingdom. Tel: +44.207.424.4422; Fax: +44 207 424 4433; E-mail: gr.davies@elsevier.com

PATIENT RESOURCES

None available

NGC STATUS

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