This syllabus is a guide for candidates wishing to pursue certification in congenital heart echocardiography. The candidate is expected to know and be skilled in basic principles of cardiac ultrasound. Additionally, they should know and understand the range of congenital heart defects in children and adults, and acquired heart defects in childhood. Although not all candidates will have the opportunity to train or work in cardiac surgical centres, they should be competent in the echocardiographic evaluation of unoperated congenital heart disease, the assessment of patients for surgical or catheter treatment, and post-procedural review. Intraoperative and cardiac catheter guidance for interventions is more advanced and the candidate is not expected to have experience of this, although an understanding of this is encouraged.

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CONGENITAL HEART DISEASE ECHOCARDIOGRAPHY

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1. GENERAL CONCEPTS

1.1. The clinical role of echocardiography and Doppler in paediatric and congenital heart disease
   1/ Indications for echocardiography
   2/ Information that echocardiography can and cannot provide
   3/ ‘Ruling out’ pathology (sensitivity, specificity & Baye’s theorem)
   4/ Likelihood of findings influencing patient management
   5/ Role of multimodality imaging
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      ▪ Multislice CT scan
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   2/ Specific requirements for paediatric and congenital echocardiography laboratories
   3/ Provision and indication for specialised techniques, e.g. Transoesophageal echocardiography
   4/ Availability and access
   5/ Controlling workload
   6/ Training & motivation of staff
   7/ Audit, Quality Control, Clinical Governance

1.3. Relationship with patients
   1/ Explaining the procedure in terms relevant to the patient/parents
   2/ Respect for patients’ dignity and cultural backgrounds
   3/ Relationships with patient, parents and colleagues
   4/ Handling requests for information about the study findings

1.4. Conscious sedation in children
   1/ Explaining the procedure in terms relevant to the patient/parents
   2/ Specific environment for performing studies in children/adults with CHD
   3/ Indications for conscious sedation
   4/ Precautions, dosage, follow-up
1.5. Reporting and documentation of paediatric and congenital studies

1/ Standard methods and terminology used for describing congenital heart disease (segmental sequential analysis)
2/ Distinction between technical and clinical reports
3/ Responsibility for reporting
4/ Medico-legal considerations (Data Protection Act)

1.6. Safety of ultrasound

1/ Potential hazardous biological effects
   ▪ Heating, resonance and cavitation effects
2/ Measurement of beam intensity (Spatial Peak Temporal Average, [SPTA])
3/ Practical precautions: power levels, use of colour and continuous wave Doppler

2. IMAGING PHYSICS AND INSTRUMENTATION

2.1. Concepts and terminology of cardiac ultrasound

1/ Concept of compression waves
2/ Definitions: frequency, wavelength, propagation velocity
3/ Units of measurement: Hz and MHz, Decibel
4/ Comparison of Ultrasound with audible sound

2.2. Propagation of ultrasound through tissues

1/ Speed of sound in different body tissues
2/ Frequency range used for diagnostic imaging in children and adults with CHD
3/ Distinction between specular reflection and backscatter
4/ Principles of attenuation and scattering

2.3. Ultrasound Transducers

1/ Piezo-electric effect
2/ General concepts of transducer construction
3/ Characteristics of the ultrasound beam
   ▪ Near (Fresnel) & Far (Fraunhofer) zones, side lobes
4/ Beam steering methods: mechanical & electronic
5/ Focusing methods, including multiple transmit focusing

2.4. Imaging physics

1/ Factors affecting choice of imaging frequency
   ▪ Typical practical values for adults & children
2/ Measurement and optimisation of resolution: axial, azimuthal and elevation
3/ Broad-band imaging
4/ Harmonic imaging
5/ B mode and M Mode methods
6/ Scanning speed limitations, relationships between pulse repetition frequency, frame rate, lines per frame, field of view, depth to be imaged
7/ Concept of Parallel Processing and its influence on frame rate and image quality
8/ Effect on evaluation of rapid motion, temporal resolution
9/ Grey scale and dynamic range
10/ Lateral resolution and grating artefacts
11/ Reverberation artefacts
12/ Limiting factors for detecting small targets

2.5. Echo Instrumentation
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   ▪ Transmit power; overall gain; time gain compensation; reject, logarithmic compression
2/ Signal processing, dynamic range, pre-processing, post processing
3/ Optimisation of imaging parameters including
   ▪ Transducer frequency, scan angle, gamma correction, spatial and temporal smoothing

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   ▪ Parasternal, apical (4, 5 and 2-chamber), subcostal, suprasternal, right parasternal, long and short axis
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2/ Determination of volumetric flow
3/ Continuity equation
4/ Laminar & turbulent flow: Reynolds’ equation (qualitative)
5/ Transition from Laminar to turbulent flow: inlet jet
6/ Bernoulli equation

3.2. Basic Principles of Doppler
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2/ The Doppler equation: factors influencing magnitude of Doppler shift
3/ Spectral analysis: fast Fourier transform (qualitative)
4/ The spectral Doppler display: determination of mean, modal and peak velocities
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6/ The effect of beam angle errors on Doppler velocities
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   ▪ Influence on aliasing of: transducer frequency; sample depth (range x velocity product); and beam angle
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2/ The ‘stand-alone’ Doppler probe (pencil probe)
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6/ Representation of signal strength by image intensity
7/ How aliasing manifests on the spectral display

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1/ On-screen measurement of length, slope, area, volume and time interval, and their significance for 2-D images, M-mode and spectral Doppler displays
2/ Standard M-mode measurements and calculations
   ▪ Shortening Fraction
   ▪ TAPSE
   ▪ Wall thickness
3/ Limitations of measurements and/or calculations
4/ Peak and Mean pressure gradient measurements by Doppler and their relationship to catheterisation data

3.6. Tissue Doppler Imaging (TDI) and Deformation
1/ Understand principles of TDI
2/ Identify a’, e’ s’ on PW-TDI
3/ Identify a’, e’ s’ on PW-TDI
4/ Understand age and size related differences in TDI measurements

3.7. Understand concept of Displacement imaging • Strain and strain rate
1/ Definition
2/ Direction of deformation

3.8. Speckle Tracking Echocardiography
1/ Principles
2/ Physical origin of speckles
3/ Tracking of speckle motion

3.9. Clinical applications
1/ Haemodynamic assessment - Assessment of filling pressures
2/ Systolic (global and regional) function / Diastolic function
3/ Detection of subclinical myocardial dysfunction

4. CARDIAC ANATOMY AND PHYSIOLOGY FOR PAEDIATRIC AND ADULT CONGENITAL ECHOCARDIOGRAPHY ANATOMY OF THE TORAX

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2/ Lungs & pleura
3/ Heart & pericardium
4/ Mediastinum
5/ Blood vessels within the thorax

4.2. General concepts of cardiac morphology and echo identification
1/ Atrial situs
   ▪ Definition, abdominal aorta and great vein relationship
2/ Systemic venous return: morphology
3/ Pulmonary venous return: morphology
4/ Atrial anatomy
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   ▪ Ventricular shape
   ▪ Right-handed vs. left handed ventricular configuration
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7/ Semilunar valves: anatomy of pulmonary and aortic valve
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  • Situs inversus
  • Right and left isomerism
2/ Atrioventricular connections
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  • Discordant
  • Double inlet
  • Absent connection
  • Straddling valves
  • Criss-cross connections
3/ ‘Univentricular’ heart: description of different variants
4/ Ventriculoarterial connections
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  • Discordant
  • Single outlet
  • Double inlet
5/ Great artery relationships

4.4. The cardiac cycle
1/ Temporal relationships of the ECG, chamber pressures and valve movements
2/ Typical values for intracardiac pressures
3/ Relationship of valve movements to heart sounds

4.5. Fetal and neonatal physiology
1/ The fetal circulation: how it differs from the postnatal circulation
2/ Circulatory changes at birth: the neonatal circulation
3/ Adaptations in circulatory physiology during the first weeks of life

4.6. The physiology of congenital heart disease
1/ Causes of chamber dilation and hypertrophy
2/ Ventricular pressure and volume overload
3/ Physiological effect of shunts at atrial, ventricular and great artery level
4/ Physiological effect of regurgitation through all four valves
5/ Physiological effect of stenosis on all four valves

5. THE DIFFERENT CONGENITAL HEART DEFECTS AND TREATMENT FOR THE PAEDIATRIC AND CONGENITAL ECHOCARDIOGRAPHER

5.1. Septation defects
1/ Ventricular septal defect (VSD)
   • Assessment of unoperated VSD
     - Location and size
     - M-mode and 2D features
     - Assessment of shunt size and pulmonary pressure
     - Anterior or posterior malalignment
   • Associated features
     - Aortic valve cusp prolapsed
     - Discrete subvalvular aortic stenosis
     - Double chambered right ventricle
   • Echocardiographic assessment of surgical treatment for VSD
     - Post-operative complications: e.g. residual defects, sub-aortic stenosis
   • Echocardiographic assessment of percutaneous treatment for different types of VSD

2/ Atrial septal defect (ASD)
   • Assessment of unoperated ASD
     - Secundum, primum and sinus venosus defect
     - Location and size
     - M-mode and 2D echo features
     - Assessment of shunt size
     - Common associated lesions
   • Other causes of right ventricular volume overload
   • Echocardiographic assessment of surgical treatment of different types of ASD
Echocardiographic assessment of percutaneous treatment of ASD

3/ Atrioventricular septal defect (AVSD)
- Assessment of unoperated AVSD
  - Size of atrial and ventricular components
  - Atrioventricular valve function
  - Ventricular imbalance
  - Ventricular imbalance
- Echocardiographic assessment of post-operative AVSD
  - Atrioventricular valve function
  - Sub-aortic stenosis
  - Residual defects

5.2. Shunt lesions not caused by septation defects
1/ Arterial duct
- Assessment of patent arterial duct
  - Imaging planes for isolated duct
  - Assessment of shunt size
  - Ductal flow patterns and pulmonary artery pressure
- Anatomical variations with different cardiac defects
- Other defects causing shunt at great artery level
- Echocardiographic assessment of surgical and percutaneous treatment for patent arterial duct

2/ Partial anomalous pulmonary venous drainage (PAPVD)
- Assessment of partial anomalous pulmonary venous drainage
  - Anatomy – which veins drain where
  - Associated defects
  - Haemodynamic effect
- Echocardiographic assessment of surgical treatment for PAPVD

3/ Basic anatomy and echocardiographic features of other acyanotic shunts
- Aortopulmonary window
- Unroofed coronary sinus
- Origin of one pulmonary artery from aorta
- Sinus of Valsalva fistula

5.3. Cyanotic congenital heart defects
1/ Ventriculoarterial discordance (transposition of the great arteries)
- Assessment of unrepaired ventriculoarterial discordance
Anatomy
- Coronary artery anatomy
- Associated features: VSD, pulmonary stenosis, coarctation

**Echocardiographic assessment of surgical treatment for ventriculoarterial discordance**
- Atrial switch (Mustard, Senning)
- Arterial switch
- Rastelli procedure

2/ Tetralogy of Fallot (TOF)/pulmonary atresia with ventricular septal defect (PA/VSD)

- Assessment of unrepaired TOF/pulmonary atresia with VSD
  - Anatomy
  - Assessment of VSD
  - Right ventricular outflow tract assessment
  - Coronary artery anatomy
  - Sources of pulmonary blood flow
  - Size of branch pulmonary arteries
  - Arch laterality

**Echocardiographic assessment of surgical treatment for TOF/pulmonary atresia with VSD**
- Palliations: arterial shunt, right ventricular outflow tract stent, arterial ductal stent, pulmonary valve balloon
- Valve sparing
- Transannular patch
- Right ventricle to pulmonary artery conduit
- Haemodynamic effect of residual stenosis or regurgitation

3/ Pulmonary atresia with intact septum (PA/IVS)

- Assessment of unrepaired PA/IVS
  - Anatomy
  - Pulmonary valve morphology
  - Coronary artery anatomy/fistulae/sinusoids
  - Right ventricular morphology

**Echocardiographic assessment of percutaneous and surgical treatments for PA/IVS**
- Arterial shunt, right ventricular outflow tract stent, arterial ductal stent, radiofrequency perforation and pulmonary valve balloon

4/ Total anomalous pulmonary venous drainage (TAPVD)

- Assessment of unrepaired TAPVD
  - Anatomy
  - Location: supracardiac, cardiac, infracardiac
  - Size of pulmonary veins
- Associated features
  - Echocardiographic assessment of surgical treatment for TAPVD
    - Patency of confluence
    - Pulmonary vein stenosis
    - Right heart pressures

5.4. Other complex lesions

1/ Common arterial trunk (CAT)
  - Assessment of unrepaired CAT
    - Anatomy
    - Morphology and function of truncal valve
    - Size and position of VSD
    - Commital of truncal valve to ventricle
    - Origin of pulmonary arteries
    - Coronary artery origins
    - Associated defects
  - Echocardiographic assessment of surgical treatment for CAT
    - Truncal valve stenosis/regurgitation
    - Pulmonary artery stenosis
    - Right ventricle to pulmonary artery conduit function

2/ Lesions with single ventricle physiology
  - Different variants: double inlet ventricles, double outlet ventricles, hypoplastic/absent atroventricular connections etc.
  - Anatomy
    - Morphology of ventricle
    - Source of pulmonary blood flow
    - Source of systemic blood flow
    - Atrioventricular and ventriculoarterial valve function
  - Echocardiographic evaluation after staged palliation for single ventricle physiology
    - Pulmonary artery banding
    - Norwood procedure
    - Arterial shunt
    - Bidirectional Glenn/hemi-Fontan
    - Fontan/total cavopulmonary connection (TCPC)
  - Echocardiographic evaluation of the Fontan/(TCPC)
    - Types of Fontan/TCPC (atriopulmonary, lateral tunnel, extracardiac conduit)
    - Obstructions to the Fontan tunnel
    - Thromboses
    - Single ventricle function
    - Fenestration flow
3/Atrioventricular and ventriculoarterial discordance (AV/VA discordance)
- Assessment of unrepaired AV/VA discordance
  - Ventricular morphology
  - Associated defects
- Echocardiographic follow-up of the unoperated patient
  - Function of the systemic right ventricle and tricuspid valve
- Echocardiographic follow-up of the operated patient
  - Double switch (atrial and arterial switch)

4/Double outlet right ventricle (DORV)
- Assessment of the unoperated patient with DORV
  - Arrangement of the great vessels (and the relevant physiology)
  - Size and location of the VSD
  - Associated lesions
- Echocardiographic assessment of the surgical treatment of DORV
  - Residual VSD
  - Left or right ventricular outflow tract obstruction

5.5. Congenital valve disease
1/ The mitral valve
- Assessment of the congenitally abnormal mitral valve
  - Anatomy and different variants of mitral valve anomalies
  - Nomenclature for mitral valve leaflets
  - Description of the valve
  - Description of subvalvar apparatus
  - Measurement of orifice area by planimetry
  - Assessment of severity of stenosis/regurgitation
- Doppler assessment of the mitral valve
  - Mean and end-diastolic gradient
  - Area by ‘pressure half-time’: technique and limitations
- Mitral valve prolapse: definition and echocardiographic aspects
- Echocardiographic assessment of surgical mitral valve repair

2/The aortic valve
- Assessment of the congenitally abnormal aortic valve
  - Morphology of the aortic valve
  - Number of leaflets
- Assessment of the left ventricle: size, hypertrophy, systolic and diastolic function
- Associated left ventricular outflow tract abnormalities
- Effect on the aortic root
- Assessment of severity of stenosis/regurgitation

- **Doppler assessment of the aortic valve**
  - Peak and mean gradients
  - Apical, right parasternal and suprasternal positions
  - Continuity equation

- **Echocardiographic assessment of surgical and percutaneous treatments for congenital aortic valve disease**
  - Aortic valve balloon
  - Aortic valve repair
  - Aortic valve replacement
  - Ross procedure

3/ The pulmonary valve

- **Assessment of the congenitally abnormal pulmonary valve**
  - Morphology of the pulmonary valve
  - Number of leaflets
  - Assessment of the right ventricle: size, hypertrophy, systolic and diastolic function
  - Associated right ventricular outflow tract abnormalities
  - Assessment of severity of stenosis/regurgitation

- **Echocardiographic assessment of surgical and percutaneous treatments for congenital pulmonary valve disease**
  - Pulmonary valve balloon
  - Pulmonary valve repair
  - Pulmonary valve replacement
  - Right ventricular remodelling

4/ The tricuspid valve (including Ebstein’s Malformation)

- **Assessment of the congenitally abnormal tricuspid valve**
  - Morphology of the tricuspid valve
  - Position of tricuspid valve annulus
  - Number of leaflets
  - Assessment of the right ventricle and right atrium: size, function
  - Associated abnormalities
  - Assessment of stenosis/regurgitation
Echocardiographic assessment of surgical and percutaneous treatments for congenital tricuspid valve disease
- Tricuspid valve repair
- Cone repair

5/Prosthetic Valves
- 2D, M-Mode and Doppler features of the main types of replacement valves
  - Ball & cage
  - Tilting Disc
  - Bi-leaflet
  - Stented Bioprostheses
- Age-related deterioration of bioprostheses
- Understand role of TOE in examining normal and malfunctioning prosthetic valves
- Prosthetic valve stenosis
  - Assessment by 2D, M-mode and Doppler
  - Normal ranges
  - Use of Continuity Equation for aortic prostheses
- Prosthetic valve regurgitation
  - Trans-versus para-valvar regurgitation
  - Normal versus abnormal regurgitation
- Assessment by CW, PW and Colour Doppler
- Colour artefacts from mechanical prostheses

5.6. Left outflow obstruction
1/ Subvalvular/supravalvar aortic stenosis
- Anatomy and variants
- Associated lesions
- Echocardiographic assessment of severity
- Evaluation of surgical treatment

2/ Coarctation of the aorta
- Type of narrowing
- Site of narrowing
- Morphology and anatomy of the whole arch
- Associated defects
- Echocardiographic assessment
  - Different echocardiographic appearances
  - Spectral Doppler appearances
  - Peak velocity and diastolic pattern
  - Relationship to measured gradient
- Relationship to severity of obstruction
  ▪ Effect of arterial duct on imaging and Doppler appearances
  ▪ Echocardiographic assessment of percutaneous and surgical
treatment of coarctation of the aorta
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    - Left ventricular remodelling

3/Interrupted aortic arch
  ▪ Site of interruption
  ▪ Associated lesions
  ▪ Echocardiographic assessment of surgical treatment of
interrupted aortic arch

5.7. Right outflow obstruction
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    ▪ Assessment of surgical treatment for anomalous origin of the
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    ▪ Physiological effect
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for coronary fistulae

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  2/Intracardiac Masses
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    ▪ Echocardiographic features intracardiac tumours
    ▪ Differentiation of cardiac tumours
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   ▪ Echocardiographic features of the normal aortic root, sinuses of Valsalva, ascending aorta and aortic arch
   ▪ 2-D, M-mode and Doppler features of
     - Marfan’s syndrome
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4/Pericardial Disease
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     - Relationships of serous pericardium to heart and great vessels
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     - Differentiation from pleural effusion
     - Assessment of volume of pericardial fluid
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     - Effect on IVC
     - Effect on A-V valve flow velocities
   ▪ Features of pericardial constriction
     - Effect on A-V valve flow velocities
     - Effect of respiration
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     - Differentiation from restrictive cardiomyopathy

6. ACQUIRED HEART DISEASE

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   2/ Echocardiographic follow-up

6.2. Infective Endocarditis
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2/ Preferred locations for vegetations
    ‘Jet’ lesions
    Congenitally abnormal structures
    Hypertrophic cardiomyopathy
3/ Complications: abscess, fistula, perforation
4/ Understand role of TOE in suspected endocarditis
5/ Echocardiographic follow-up

6.3. Rheumatic fever
   1/ Common echocardiographic features
   2/ Evaluation of severity
   3/ Echocardiographic follow-up

6.4. Intravascular thrombosis
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   2/ Echocardiographic follow-up

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       Methods of measuring LV volume, including biplane area, area-length Simpson’s rule methods
       Echocardiographic calculation of cardiac output
   2/ Diastolic function
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7.2. Pulmonary hypertension
   1/ Pulmonary Hypertension
       2-D, M-mode and Doppler features of pulmonary hypertension
       Causes of pulmonary hypertension
         - Idiopathic
         - Related to congenital heart disease
         - Chronic pulmonary emboli
         - Secondary to lung diseases
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         - Medication related
       Measurement of pulmonary pressures from tricuspid and pulmonary regurgitant flow velocities
       Assessment of inferior vena cava contraction

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1/ Dilated Cardiomyopathy
- 2D, M-mode and Doppler features of dilated cardiomyopathy
- Detection and assessment of associated lesions: functional valve regurgitation
- Thrombus in cardiac chambers
- Pericardial effusions
- Associated abnormalities, e.g. mitral regurgitation
- Role of echocardiography in assessment and follow-up

2/ Hypertrophic Cardiomyopathy
- 2D, M-mode and Doppler features of hypertrophic cardiomyopathy
- Differentiation from other causes of hypertrophy, e.g. ‘athletic heart’
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- Assessment of right ventricular involvement
- Associated abnormalities, e.g. mitral regurgitation
- Role of echocardiography in assessment and follow-up

3/ Restrictive Cardiomyopathy
- 2D, M-mode and Doppler features of restrictive cardiomyopathy
- Differentiation from pericardial constriction
- Techniques for measurement of diastolic dysfunction
- Associated abnormalities, e.g. mitral regurgitation
- Role of echocardiography in assessment and follow-up