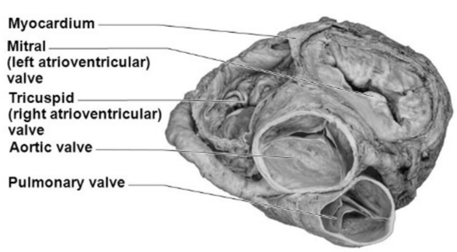
Congenital valvular heart disease

Dr Edgar Tay
Director of Structural Heart Service
National University Heart Centre
Echo Singapore 2018



Least studied valve?

- Least accessible echocardiographic windows
- Mostly congenital in etiology
- Increased number of ACHD patients and availability of Rx
- Hemodynamic significance

Classification of major diseases that affect the pulmonary valve

Pulmonary atresia	Pulmonary stenosis	Pulmonary regurgitation	Mixed PS and PR
With intact ventricular septum a/w: • hypoplastic RV and tricuspid valve • ASD • Coronary anomalies	**Valvular**: Dome shaped/Dysplastic (part of Noonans) Bicuspid/Quadricuspid Acquired	Physiologic: trivial to mild	Carcinoid
With VSD (a form of tetralogy of fallot associated with MAPCAs	**Supravalvular**: Congenital a/w: A) Alagille syndrome B) Keutel syndrome C) Congenital rubella D) TOF E) William syndrome Acquired /iatrogenic: Takayasu	Primary: Some forms of TOF (eg Absent pulm valve) Acquired causes Post operative/Post valvuloplasty	Rheumatic
	Subvalvular: Primary infundibular stenosiss DCRV Secondary infundibular hypertrophy	Acquired: PAH	Infectious/iatrogenic

Focus on the pulmonary valve

- Pulmonary valve
  - Stenosis
  - Regurgitation

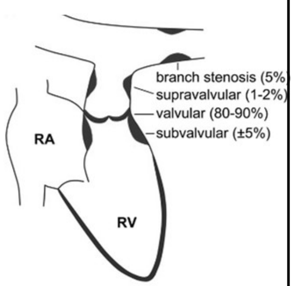
Valvular pulmonary stenosis

- 7% of children born with CHD
- Associations with dilated PA
- Noonans: Hypoplastic pulmonary annulus and supravalve narrowing

Differentials

- **Infundibular stenosis** (This is associated with TOFs)
- Should be suspected when there is increased RV thickness and dynamic late peaking doppler systolic signals in subvalvular region
- Also different from **Double Chamber RV**: Here a fibromuscular collar develops between the RV inflow and RV outflow tract. Short Axis view probably best to show this (with RVOT in plane)
- **Supravalvular stenosis** (uncommon and associated with Willams Syndrome/TOF, congenital Rubella syndrome

King et al. Curr Cardio Rep 2015



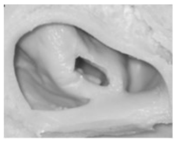
Hoffman et al. Heart 2004

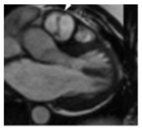
Echocardiographic assessment

- Morphology of valve
- Hemodynamics
- Right ventricle

Morphology of valve

- Thickening
- Commisural fusion
- Common morphologies – Bicuspid, dome shaped(40-60%) or dysplastic (20%)
- Annular size



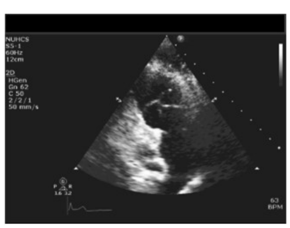


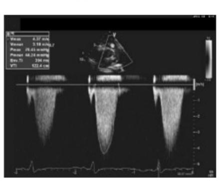
Arain et al. Ann Paed cardiol 2012
Kovalchin et al JACC 1997

Hemodynamics

- Assessment of pulm valve from parasternal short axis image or apical four chamber with anterior tilt (CW)
- Or TR velocity
- Good correlation with peak to peak gradients on invasive catheterization

Cantinotti et al. Echocardiography 2017





Severity of Pulmonary Stenosis

Mild	Peak gradient <36mmHg, peak velocity <3m/s)
Moderate	Peak Gradient 36-64mmHg, peak velocity 3-4m/s
Severe	Peak gradient > 64mmHg, Peak velocity > 4m/s; Mean gradient >35mmHg

RVSP assessed by TR velocity

Adapted from Stout et al. Circulation 2018

Right ventricle

- Right ventricular hypertrophy
- Assessment of right ventricular systolic or diastolic function (TAPSE/TDI) (*differences in adult and children*)

Koestenberger et al. Eur heart J Cardiovasc Imaging 2014
Koestenberger et al. Am J Cardiol 2012

Recommended follow up

Frequency of Routine F/U and testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Clinical review	36-60	24	6-12	2-6
ECG	36-60	24	12	12
TTE	36-60	24	12	12
Exercise test	As needed	24	24	12

Adapted from Stout et al. Circulation 2018

Management

- Those with mild PS have a very good natural history and intervention usually unnecessary
- Mod/severe PS may require intervention:
  - Surgical valvotomy
  - Balloon valvuloplasty
  - Indications: Peak transvalvular gradient >60mmHg (or >/=50mmHg with symptoms; or mean of 40mmHg or >30mmHg with symptoms)

Pulmonary regurgitation

- Important in ACHD due to significant patients who have survived to adulthood following TOF repair/those who have had balloon valvuloplasty
- Severe PR is associated with RV dysfunction, arrhythmias, heart failure and mortality
- Trace to mild PR is common and probably has minimal hemodynamic consequence

Anatomic considerations

- Semilunar with 3 cusps, thinner than aortic valve and its plane is orthogonal to AV
- Lack fibrous continuity with tricuspid valve
- Anterior structure so more difficult to image
- Limited windows for both TTE and TEE

Etiology

- Can occur in native congenital heart disease or following intervention (eg TOF repair or balloon pulmonary valvuloplasty for PS)
- Differentials (aquired disease generally rare), functional PR usually smaller in volume and leaflets normal

Morphology

- Bicuspid/Quadricuspid (no. of leaflets)
- Doming/Prolapse (motion)
- Hypoplastic, dysplastic or absence (Structure)

Echocardiographic assessment of valve hemodynamics

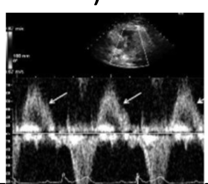
- Colour flow doppler
- Pulsed and continous wave doppler
- Quantitative doppler

Parameters	Mild	Moderate	Severe
Qualitative			
Pulm valve morphology	Normal	Normal/Abnormal	Abnormal
Colour flow PR jet width	Small, usually <10mm in jet length with narrow origin	Intermediate	Large with wide origin, may be brief in duration
Reversal flow in pulm arteries	Absent	Absent	Present
CW signal of PR jet	Faint/slow deceleration	Dense/variable	Dense with steep deceleration, early flow termination
Pulm vs aortic flow by PW	Normal/slightly increased	Intermediate	Greatly increased
Semiquantitative			
VC width	Not defined	Not defined	Not defined
Pressure half time	Not defined	Not defined	<100ms
Jet width ratio	Not defined	Not defined	50-65%
Quantitative			
ERO mm^2	Not defined	Not defined	Not defined
R Vol (ml)	Not defined	Not defined	Not defined
RV size			

Lancellotti et al. EHJ CV imaging 2013

Doppler (Pulse wave)

- Align insonation beam in the RPA and LPA
- Obtain pulse wave doppler from both the branch Pas (if main PA used specificity drops to 39%)

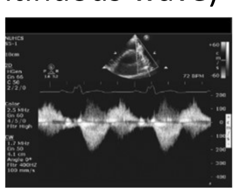


Advantage
Simple supportive sign

Depends on compliance of the PA
Brief velocity reversal is normal

Colour Doppler (Continuous wave)

- Align insonation beam with the flow
- PSAx view or subcostal view

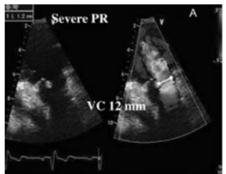


Advantages
- Simple
- Density is proportional to the number of RBC reflecting the signal
- Faint or incomplete jet – mild PR

Qualitative
Central jets may appear more dense then eccentric jets
Difficult to differentiate moderate from severe

Vena contracta

- Zoom the parasternal short axis/subcostal view
- Visualize proximal flow convergence distal jet and narrow neck in a single view
- Measure in diastole below PV



Advantages
- Surrogate for ERO
- Independence of flow rate and driving pressure for a fixed orifice
- Less dependence on technical factors

Difficult with multiple jets
Eccentric jets
Not well validated

Vena contracta width to annular diameter ratio

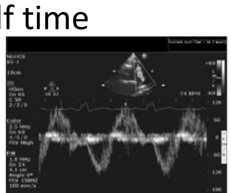
- Zoom the parasternal short axis/subcostal view
- Visualize proximal flow convergence distal jet and narrow neck in a single view
- Measure in diastole below PV

Advantages
- Simple and sensitive screen for PR
- Rapid quantitative assessment

Underestimates in eccentric jets
Overestimates in central jets
PR jet may expand unpredictably below PV
Requires clear assessment of annulus

Pressure half time

- Align insonation beam with the flow
- Parasternal short axis or subcostal view



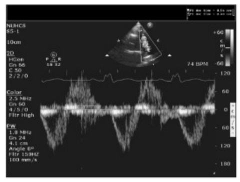
Advantages
- Simple
- Specific sign of pressure equalization
- Values<100ms consistent with severe PR

Poor alignment may result in eccentric jets showing PHT<100ms
Affected by RV and PA pressure differences (eg diastolic RV dysfunction)

Silversides et al. JASE 2003

PR index

- Align insonation beam with the flow
- Parasternal Short Axis or Subcostal view
- Ensure complete forward and regurgitant flow spectral doppler
- <0.77 correlates well with severe PR on CMR



Advantages
- Uses combination of PR duration and diastolic duration
- Accounts for pressure difference between PA and RV

Affected by RV diastolic dysfunction and RV diastolic pressure

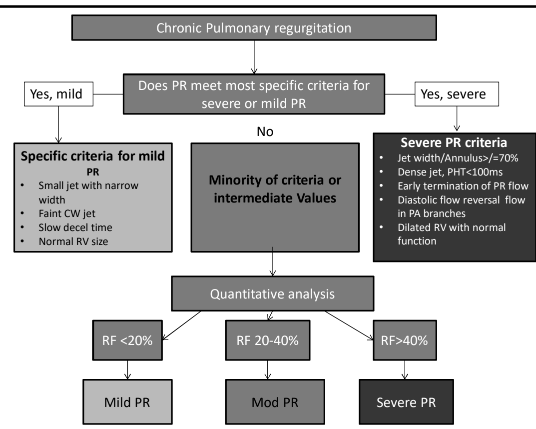
Li Wei et al. Am Heart J 2004

Quantitative doppler (Regurgitant volume and fraction)

- $RVol = SV_{RVOT} - SV_{LVOT}$
- $RF = RVol/SV_{RVOT}$
- Pulmonary annulus from PSAx measured during early ejection just below PV
- Pulse doppler in RVOT from PSAx
- Aortic annulus measured in early systole in PLAx
- Pulsed doppler in LVOT from apical views

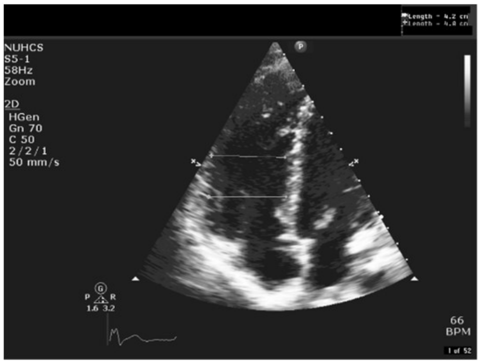
Advantages
- Quantitative, valid with multiple jets and eccentric jets

RVOT diameter not always easily measured
ERO not validated
If AR present may need to use Mitral annulus
Little experience



Zoghbi et al. JASE 2017

Right ventricle



Right ventricle

- Right ventricle dilatation (global or regional)
  - Mild PR (usually RV normal in size)
  - Acute severe PR (also normal in size)
  - "normal" = Mid RV dimension ≤33mm,RVEDA ≤28cm^2, RVESA ≤16cm^2
- Right ventricular function(systolic/diastolic):
  - FAC (Low to modest)
  - TAPSE (weak correlation)
  - TDI (fair when infundibular EF better)
  - Diastolic function – few studies

Valente et al. JASE 2014

Conclusion

- The pulmonary valve has been neglected but is becoming increasingly important due to the increase in patients with ACHD and procedures done
- Most pulmonary valve diseases are of congenital etiology
- Complete assessment of the pulmonary valve requires the evaluation and reporting of its morphology, hemodynamics as well as its impact on the right ventricle