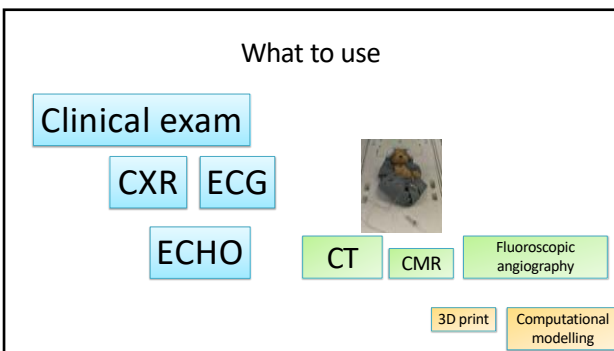




1



2



3

Prior to initial assessment

- Post-natal or antenatal diagnosis
- Gestation and birth weight
 - sepsis
 - Other congenital anomalies
- Review of antenatal notes
 - RV dysfunction
 - Tricuspid regurgitation
 - Atrial septal restriction: ?lymphangiectasia

4

Useful reference

European Journal of Cardio-Thoracic Surgery 38 (2022) 414–419
doi:10.1093/ejcts/ezab188

GUIDELINES

Cite this article as: Alphonso N, Angeli A, Baron O, Bellahem-Rouel H, Blum NK, Brown K et al. Guidelines for the management of neonates and infants with hypoplastic left heart syndrome: The European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Pediatric and Congenital Cardiology (AEPCC) Hypoplastic Left Heart Syndrome Guidelines Task Force. Eur J Cardiothorac Surg 2022;55:414–19.

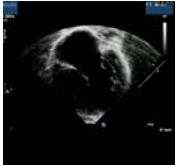
Guidelines for the management of neonates and infants with hypoplastic left heart syndrome: The European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Pediatric and Congenital Cardiology (AEPCC) Hypoplastic Left Heart Syndrome Guidelines Task Force

Nelson Alphonso^a, Annalicia Angeli^{b,c}, David J. Barron^d, Hannah Bellahem-Rouel^e, Nico A. Blom^f, Katherine Brown^g, Deborah Davis^h, Daniel Duncanⁱ, Mamy Fedigo^j, Lorenzo Gallati^k, David Hehl^l, Ulrike Herberg^m, Jeffrey P. Jacobsⁿ, Katarzyna Januszewska^o, Tom R. Karl^{p,q} (Chairman HLHS Guidelines Task Force), Edward Malec^r, Bohdan Maruszewski^s, James Montgomery^t, Christian Pizzano^{u,v}, Dietmar Schranz^w, Amanda J. Shillingford^x and John M. Simpson^y

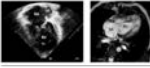
5

HLHS subtypes


Slit-like LV



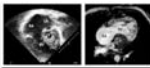
Mitral atresia/aortic atresia




Globular LV




Mitral stenosis/aortic atresia



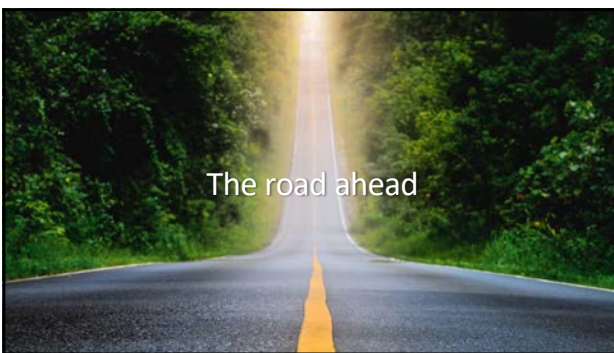
Borderline LV



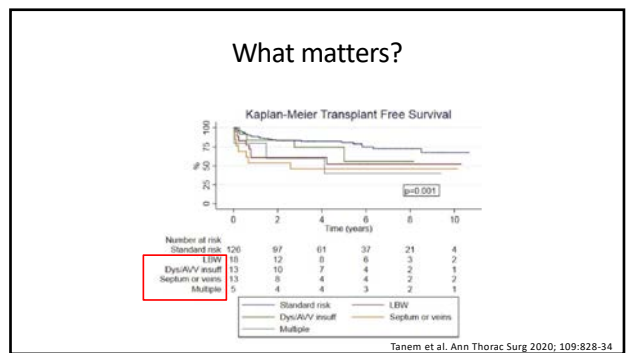
Mitral stenosis/aortic stenosis



6



7



8

What to look for– Key Points

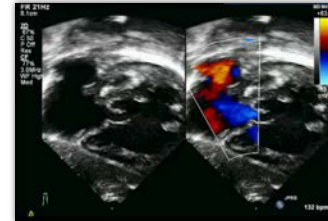
- Life maintaining structures:
 - Atrial septum
 - Arterial duct
- Tricuspid regurgitation
- Ventricular function
- Pulmonary regurgitation/stenosis
- Pulmonary artery anatomy
- Pulmonary venous abnormalities
- Coronaries
 - Fistulae
 - *Potential for use of left heart*
 - Z-scores

Alfonso et al: EJCTS 2020;58:416-499



9

Atrial communication



Unrestrictive

10

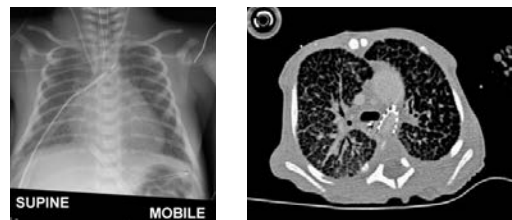
Atrial communication



Restrictive

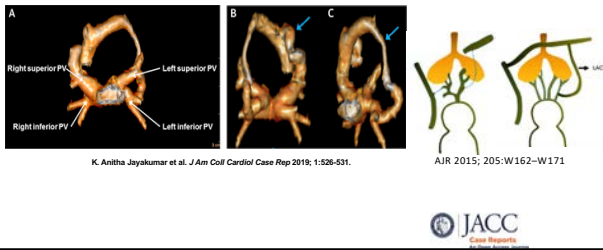
11

Pulmonary lymphangiectasia



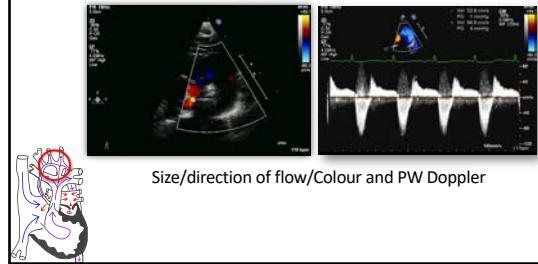
12

Levoatriocardinal vein



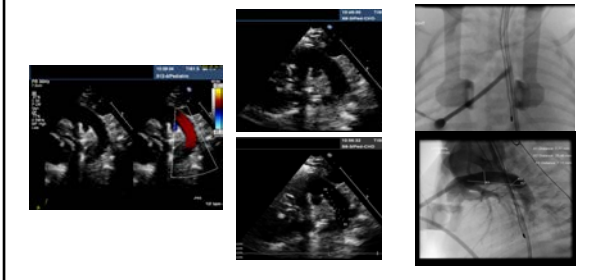
13

Arterial Duct



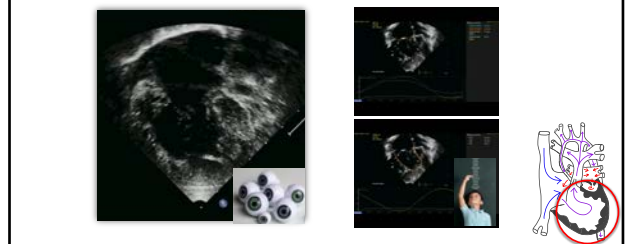
14

Arterial duct (for intervention)



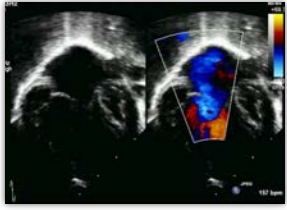
15

Right Ventricular Function

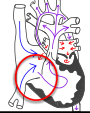


16

Tricuspid Valve

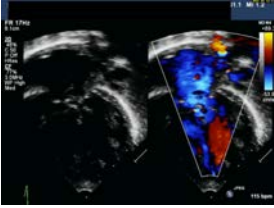
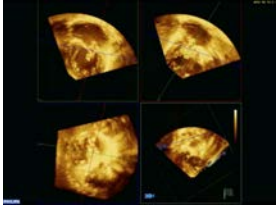


- Often abnormal in HLHS
- Assessment on colour and greyscale, 2D and 3D
- Number and position of jets
- May be exclusion factor for surgery



17

Tricuspid valve

18

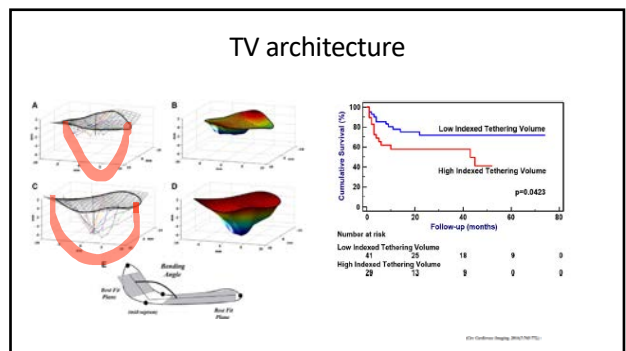
Tricuspid valve

Table 3 Primary and secondary mechanisms of regurgitation determined by surgical assessment and echocardiographic review

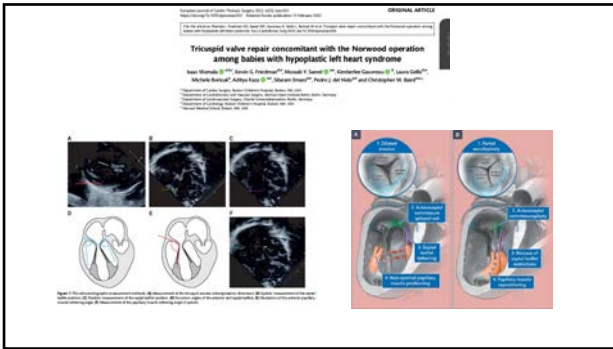
Variable	Echocardiographic review		Surgical assessment	
	Primary mechanism of regurgitation, n (%)	Secondary mechanism of regurgitation, n (%)	Primary mechanism of regurgitation, n (%)	Secondary mechanism of regurgitation, n (%)
Protrusion of leaflet	19 (59)	5 (16)	3 (9)	3 (9)
Restriction of leaflet	2 (6)	12 (38)	0	1 (3)
Annular dilatation	8 (25)	5 (16)	17 (53)	4 (13)
Leaflet dysplasia	3 (9)	5 (16)	14 (44)	12 (38)

Mechanisms of tricuspid valve regurgitation in hypoplastic left heart syndrome: a case-matched echocardiographic-surgical comparison study
 Data: Mwanali, Quaresima, Nishi, et al. *Heart Failure, Review Article*, 2020; 16(1): 1-10

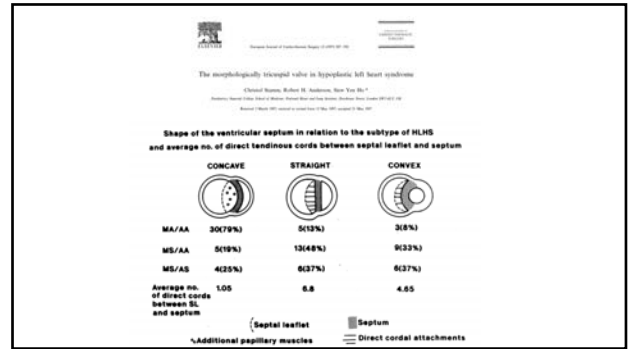
19



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21



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Pulmonary Artery/valve

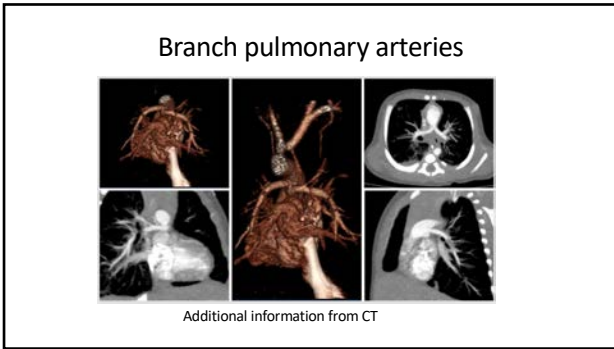
- The “new” systemic valve
- Degree of regurgitation
- Degree of stenosis, dysplasia
- Doppler, greyscale and colour assessment

23

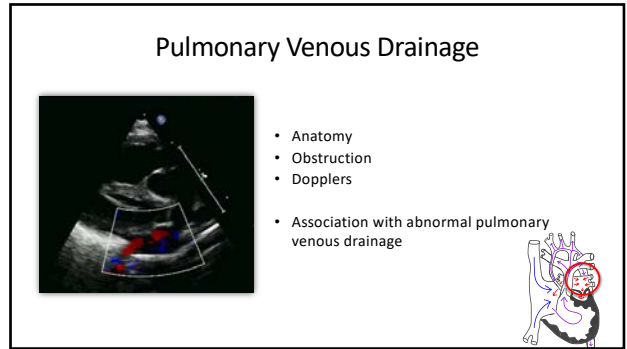
Branch Pulmonary Arteries

- Good size essential for a good surgical result at all stages
- Greyscale and colour assessment, plus Doppler

24



25



26

Abnormal pulmonary venous drainage

PEDIATRIC CARDIOLOGY

Patterns of Anomalous Pulmonary Venous Connection/Drainage in Hypoplastic Left Heart Syndrome: Diagnostic Role of Doppler Color Flow Mapping and Surgical Implications

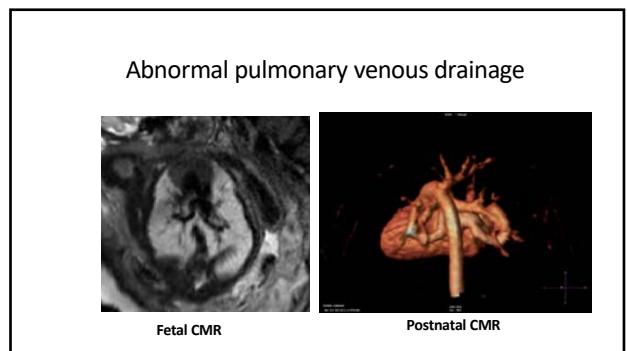
MOHAMED A. SELJEM, MD, FACC, ALVIN J. CHEN, MD, WILLIAM I. NIKPOOD, MD, PhD, FACC
Philadelphia, Pennsylvania

FROM 317 PATIENTS WITH HLHS UNDERGOING NW1 20(6.3%) had anomalous connection or drainage, or both:

- 1) Partial anomalous connection and drainage (N=2):
RPV TO RSVC , RPV TO LSVC
- 1) Total anomalous connection and drainage (N=8)
- 2) Normal connection with total anomalous drainage (N=8)
LEVOATRIOCARDINAL VEIN
- 3) Normal connection with partial anomalous drainage (N=2)
– RIGHT PV TO RIGHT OF DEVIATED SEPTUM

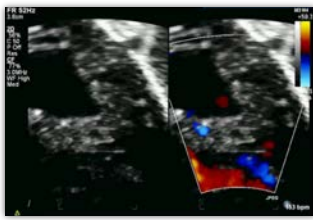
TAPVD ON FETAL CMR AT 32 WEEKS

27



28

Coronary Arteries



- Usually normal arrangement ¹
- Association with fistulae to LV: MS/AA
- Seem to be of less consequence than in PA/IVS ^{1,2}

1. Vida VL. J Thorac Cardiovasc Surg 2008;135:339-46.
2. Lloyd TR et al. Ann Heart J 1986;112:695-71.

29

Ascending Aorta and Arch

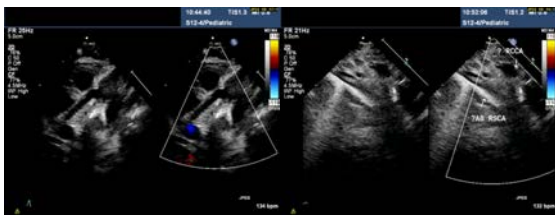


- Size of ascending aorta and arch
- Degree of hypoplasia/focal CoA
- Direction of flow in Ao arch



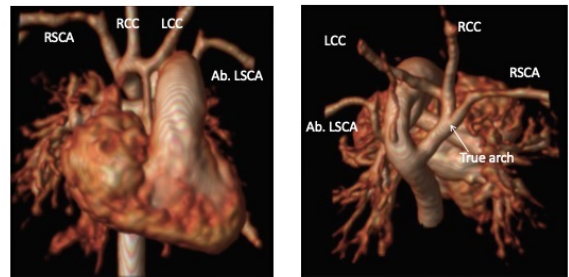
30

Echocardiography for complex arch anatomy



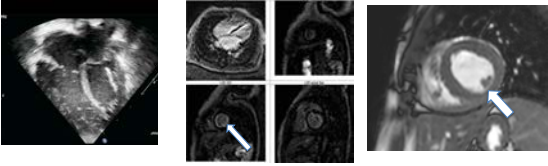
31

MRI of aortic arch



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MRI: Adequacy of LV

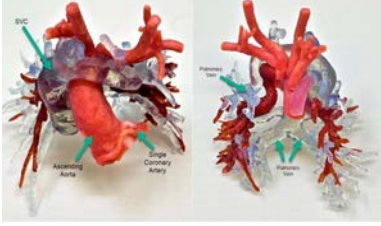


- Neonate – LV volumes
- Presence and extent of EFE ²
- Mitral Valve size and Papillary Muscle Morphology.³

1 Minich LL, et al. Am Heart J 1997;133:570-4
2 Tuo G et al. Pediatr Cardiol. 2013 Oct;34(7):1567-76
3 Velasco-Forte et al. Abstract 50389 Scientific Session 2015

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3D Printing: only for complex (non NW1)

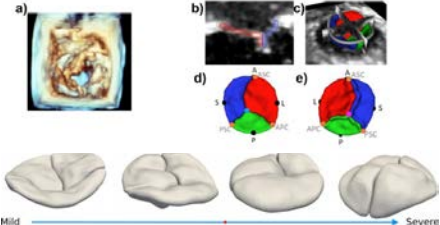


RAI, unbalanced AVSD (hypoplastic LV), TAPVD, pulmonary atresia disconnected PAs

Circ Cardiovasc Imaging. 2022;15:e014260

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Statistical shape analysis



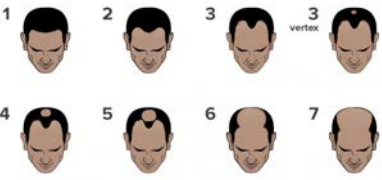
Mild → Severe

Star Abbara, Chaggar Akhondi, Heart 2022 September; 133(1): 132-140

35

CASE SELECTION/ STRATIFICATION

Norwood Scale



Norwood OT. Male pattern baldness: Classification and incidence. South Med J. 1975;68:1359-65

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What to use/when

Preoperative assessment of classical hypoplastic left heart syndrome		
Recommendations	Class	Level
Trans thoracic echocardiography is sufficient to plan initial surgery in most cases	I	C
CT or MRI should be performed because of anatomical concerns (e.g. pulmonary venous drainage path)	I	C
Chest CT for clinical suspicion of lung disease such as pulmonary lymphangiectasia	I	C
Angiography should principally be used when catheter intervention is being considered	I	C
3D echocardiography may assist in imaging the tricuspid valve to assess morphology and regurgitation	I	C
Additional features in the preoperative assessment of hypoplastic left heart complex		
The size of the left heart structures should be measured and the z-scores, calculated	I	C
Cardiac MRI may be used to define the extent of EFE	IIb	C
Predictive scores can be used to aid decision-making for SV versus bi-V circulation in critical AS	IIa	C
Predictive scores can be used to aid decision-making for SV versus bi-V circulation in unbalanced AVSD	IIa	C

Alfonso et al: EICTS 2020:58:416-499

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