Hypoplastic left heart syndrome.
Taking the long view

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Hypoplastic left heart syndrome encompasses a variety of cardiac malformations which have in common, underdevelopment or absence of left ventricle and hypoplasia of the ascending aorta.

Lev M, 1952
Noonan JA and Nadas AS, 1958
Hypoplastic left heart syndrome: 
Experience with palliative surgery 

1. Unobstructed blood flow from R.V. to systemic circulation. 
2. Reconstruction of ascending aorta and aortic arch. 
3. Restricted blood flow to pulmonary arteries. 

Norwood WI et al, Am J Cardiol 1980
FIGURE 4. Case 3. Artist’s conception of aortic atresia (top) and palliative procedure (bottom). Lower rendition illustrates ligation of patent ductus arteriosus, band on the main pulmonary artery (PA) and valved conduit between the right ventricular (RV) outflow tract and the descending aorta. Ao = aorta; ASD = atrial septal defect.

FIGURE 5. Artist’s conception of proposed physiologic repair of aortic atresia (stage 2). The main pulmonary artery (PA) and the aorta (Ao) are anastomosed using the proximal stump of the right pulmonary artery (RPA) (upper panel). The right pulmonary artery is anastomosed end to side with the superior vena cava and a valved conduit placed between the right atrium (RA) and left pulmonary artery (LPA). An interatrial baffle ensures filling of the right ventricle (Vent.) with pulmonary venous blood from the left atrium (LA). A schematic drawing of the baffle and conduit is shown in the lower panel.
Palliative reconstructive surgery for hypoplastic left heart syndrome.

Pigott JD et al, Ann Thorac Surg 1988

Ohye RG et al, NEJM 2010
Gradual decrease in hospital mortality

Modification of surgical technique
Foetal diagnosis
Pre and post-operative

The biggest challenge has been the maintenance of adequate systemic and coronary blood flow.

Right ventricle to pulmonary artery shunt in first stage palliation of hypoplastic left heart

Figure 4. After completion of aortic reconstruction and atrial septectomy, proximal anastomosis of the RV-PA shunt was performed with the heart beating.

Sano S et al,
J Thorac Cardiovasc Surg 2003
Comparison of shunt types in the Norwood procedure for single ventricle lesions.

(prospective single ventricle trial)

Ohye RG et al, NEJM 2010
Long term survival of hypoplastic left heart syndrome: meta-analysis comparing outcomes from the modified Blalock-Taussig shunt and the right ventricle to pulmonary artery shunt.

Early survival advantage of RV-PA conduit lost at 2 years then remained unchanged up to 6 years.

Cao JY et al, Int J Cardiol 2018
Change in the prevalence of risk factors for inter-stage mortality.

- RV dysfunction.
- AV valve regurgitation.
- PA hypoplasia and stenosis.

RV dysfunction may, in the long term, adversely affect the prognosis after the Fontan.
Figure 1. Regional segments:

Hill GD et al, J Am Soc Echocardiogr 2015
Circumferential indices by shunt type.

Comparison of global and regional circumferential strain and strain rate by shunt type as assessed from the short axis window.

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<thead>
<tr>
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<th>MBTS</th>
<th>RVPAS</th>
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<td></td>
<td>Mean</td>
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<tr>
<td>Circumferential strain rate</td>
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<tr>
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<td>Global</td>
<td>-1.04</td>
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</table>

Hill GD et al,
J Am Soc Echocardiogr 2015
There was no significant change in RVEF ejection fraction in the MBTS group, while the RVPAS group had significantly decreased ejection fraction compared to 14-month study (bracket).

MBTS = modified Blalock-Taussig shunt; RVEF = right ventricular ejection fraction; RVPAS = right ventricle-to-pulmonary artery shunt.
Right ventricular morphology and function following stage I palliation with a modified Blalock-Taussig shunt versus a right ventricle-to-pulmonary artery conduit.

Wong J et al, EJCTS 2016
Wong J et al, EJCTS 2016

The Athens Crossroad – 8-10 November, 2018
Pulmonary artery stenosis and hypoplasia

- Higher incidence of pulmonary artery stenosis and need for shunt intervention in RV-PA conduit group.

- RPA growth significantly greater in M.B.T.S. group.

- 94% re-operation rate on PA’s in RV-PA conduit group compared with 24% in M.B.T.S. group (Griselli et al 2006).
The long term clinical significance of these findings will become apparent by continued surveillance.
A reduction in inter-stage mortality remains a challenging issue.

a) Modification in surgical technique of right ventriculotomy and the type of position and length of RV-PA shunt.

a) Selective use of RV-PA shunt for high risk groups.

b) Inter-stage surveillance including home monitoring.
Selective use of the Blalock-Taussig shunt and right ventricle-to-pulmonary artery conduit during the Norwood procedure.

Figure 1. Actuarial transplant-free survival for RV-PA conduit and modified Blalock-Taussig shunt (mBTS) groups.

Are stem cells the next frontier for hypoplastic left heart syndrome? What are the promise, the reality, and the future?

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http://dx.doi.org/10.1016/j.jtcvs.2015.09.006
Any operative procedure being recommended to a patient should offer the best lifetime strategy.
Is there a risk to avoiding risk for younger patients with aortic valve disease?

Treasure T et al, BMJ 2011

Within a highly monitored surgical specialty, there is an obsession with peri-operative deaths that can easily be counted.
Thank You

William I. Norwood, MD

Shunji Sano, MD