Use and Outcomes of the Medical Hybrid Procedure for Stage 1 Palliation in Infants with Hypoplastic Left Heart Syndrome and Variants

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BACKGROUND AND OBJECTIVES

- Despite improvements over time, hypoplastic left heart syndrome (HLHS) and variants remain high-risk for morbidity and mortality
- Staged palliation strategies
  - Traditional surgical approach: Norwood, Glenn, Fontan
  - Hybrid Procedure: alternative strategy for initial palliation
    - Bilateral PA bands, PDA stent, +/- atrial septostomy
    - High-risk patients at most centers, routine use at a few centers
    - ~13% of patients nationally
  - **MEDICAL HYBRID (MH)**
    - Bilateral PA bands, **prostaglandins**, +/- atrial septostomy
    - Use and outcomes have not been well described
- Advantages of MH
  - Arch reconstruction may be easier without prior PDA stent
  - Avoiding PDA stent may prevent isthmus stenosis, improving retrograde coronary and cerebral perfusion
  - Possible survival benefit of inpatient interstage period?
- Disadvantages of MH
  - Prostaglandin side effects, longer hospitalization (higher cost?), central line

OBJECTIVES

1. Describe **use** of MH palliation using a large multicenter database
2. Assess **clinical outcomes** of the MH strategy compared to Stented Hybrid (SH) and Surgical Stage 1 (SS1)

METHODS

- National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) database
  - 65 centers, patients born 2016-2021, data from birth to 1 year of life
- Categorized patients into 3 groups based on **initial intervention**
  - **MH = bilateral PA bands + prostaglandins**
    - No PDA stent or surgical stage 1 prior to or simultaneous with PA bands
  - **SH = bilateral PA bands + PDA stent**
    - PDA stent prior to or simultaneous with PA bands
  - **SS1 = Norwood, DKS connection, or other appropriate surgery**
- Exclusion criteria
  - Procedure uncertain, unclear, or outside these 3 groups
  - Primary cardiac diagnosis not an indication for staged palliation
- Analyses
  - Description of MH **use**
    - Patient characteristics and risk factors, compare to SH and SS1
    - Variation across different centers
  - Analysis of MH **outcomes**
    - Subsequent procedures and clinical course
    - Survival compared to SH and SS1
  - Risk-adjusted survival analysis

RESULTS

- Initial palliation performed for 2487 patients from 65 centers
  - Excluded 64 patients
- **N = 2423 patients** from 65 centers
  - **MH group: 277 (11.4%)**
  - **SH group: 133 (5.5%)**
  - **SS1 group: 2013 (83.1%)**

Table 1. Patient Characteristics with Group Comparisons

Patient Characteristics	Total	MH	SH	SS1	*p* Value
Birth weight (kg)	3.13 ± 0.62	2.72 ± 0.72	2.95 ± 0.63	3.19 ± 0.58	**<0.001**
Gestational age (weeks)	38.4 ± 1.6	37.0 ± 2.5	37.8 ± 1.9	38.6 ± 1.3	**<0.001**
Sex male	60.4%	59.1%	60.2%	60.6%	0.95
Prenatal diagnosis	86.9%	84.7%	81.1%	87.5%	0.06
Primary cardiac diagnosis					**<0.001**
HLHS	71.9%	69.6%	57.1%	73.2%	
Other diagnosis	28.1%	30.4%	42.9%	26.8%	
Major syndrome	12.3%	21.0%	22.6%	10.4%	**<0.001**
Major non-cardiac congenital anomaly	7.7%	16.7%	11.3%	6.2%	**<0.001**
≥1/12 preop risk factors/adverse events*	45.8%	75.5%	65.4%	40.4%	**<0.001**

* **Preop risk factors/adverse events**: Acidosis, elevated lactate, elevated creatinine, periop inotrope infusion, shock (persistent or resolved), sepsis, NEC (medical or surgical), preop neurologic deficit, preop mechanical circulatory support, lifetime seizure, tracheostomy, mechanical ventilation for cardiorespiratory failure

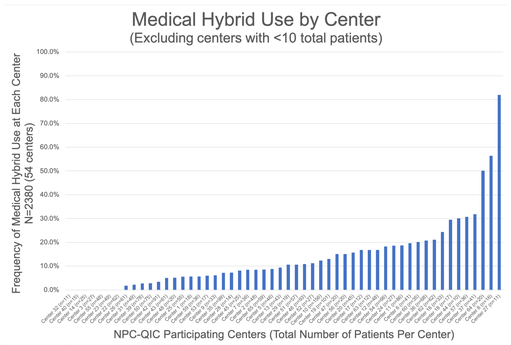

Figure 1. Frequency of MH use at each center in the NPC-QIC database

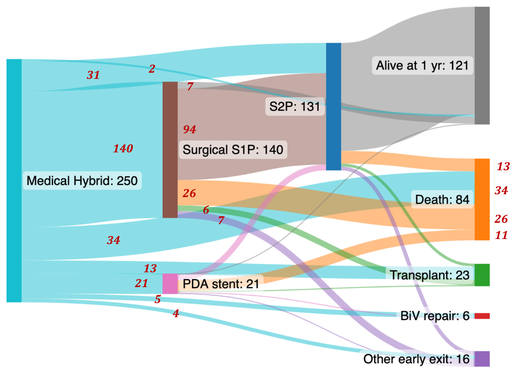

Figure 2. Sankey diagram depicting high-level clinical course of the MH patients with known outcomes. Opaque bars represent key clinical events or procedures; translucent ribbons connecting the bars represent groups of patients who progressed between the two connected events; height of each bar or ribbon is proportional to the number of patients being represented.

RESULTS

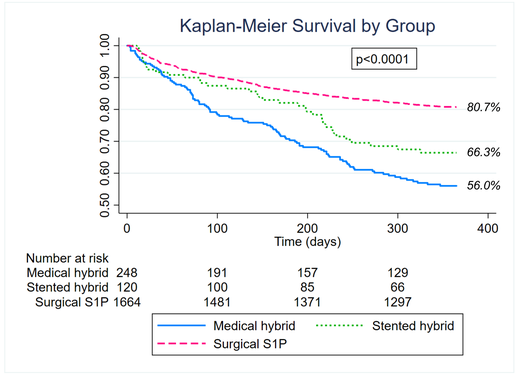

Figure 3. Kaplan-Meier one-year survival curves for the 3 treatment groups (censored at transplant or biventricular repair)

- MH preferentially used in higher-risk patients?
  - Pairwise analysis of MH vs SH, excluding single center that performed >50% SH (compare high-risk patients)
    - No significant difference in birth weight, gestational age, sex, rate of prenatal diagnosis, presence of genetic syndrome, presence of non-cardiac congenital anomaly, or presence of at least one preop risk factor

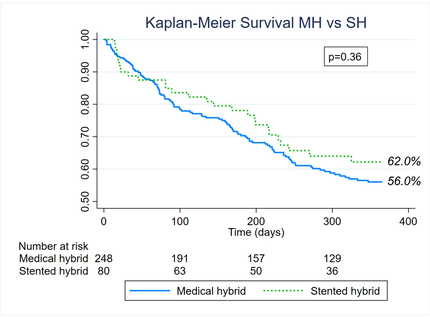

Figure 4. Kaplan-Meier one-year survival curves for MH (all) vs SH from centers that performed SH in minority of cases

- Multivariable logistic regression to elucidate predictors of MH palliation
  - Increased odds of MH:
    - ≥4 preoperative risk factors: OR 7.10 (4.59-10.97)
    - Major non-cardiac congenital anomaly: OR 1.77 (1.06-2.97)
    - Major genetic syndrome: OR 1.54 (1.05-2.26)
  - Decreased odds of MH:
    - Higher birth weight: OR 0.61 (0.48-0.77)
    - Higher gestational age: OR 0.67 (0.56-0.80)
- Cox proportional hazards regression to compare time to death/transplant while adjusting for the risk factors included in the multivariable logistic regression
  - MH had lower risk of one-year mortality/transplant compared to SS1: HR 0.54 (0.42-0.70)
  - No significant difference compared to SH: HR 0.85 (0.60-1.22)

CONCLUSIONS

- MH is used more often than SH for initial palliation of HLHS and variants
- Most patients progress from MH to subsequent surgical stage 1 procedure
- MH strategy is primarily used for high-risk patients
- Outcomes are worse than primary SS1, likely due to underlying patient risk factors
- Outcomes are similar to SH at centers that do not routinely perform SH for most patients
- Lower birth weight, lower gestational age, presence of ≥4 preoperative risk factors, presence of a major non-cardiac congenital anomaly, and presence of a major genetic syndrome are all predictors of MH palliation
- MH risk-adjusted mortality is less than SS1 and similar to SH

LIMITATIONS

- Limited information on rationale for chosen strategy
- NPC-QIC data entry was not designed with anticipation of MH palliation
- No minimum time interval between initial PA bands and later PDA stent for defining MH in this study

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