

Outcomes of Fetuses with Heart Disease Referred to a Perinatal Care Service

The Texas Children's Hospital Experience

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OBJECTIVES:

- To illustrate the experience of the Fetal Cardiology & Perinatal Pediatric Advanced Care Team (PPACT) at Texas Children's Hospital (TCH)
- To identify prenatal diagnoses at high risk for neonatal morbidity and mortality
- To describe a multidisciplinary practice involving Obstetrics, Maternal Fetal Medicine, Fetal Cardiology, Genetics, Fetal / Pediatric & Congenital Heart Surgery, Neonatology and PPACT
- To propose an “ideal” management plan for the “high risk” cardiac fetus who may benefit from PPACT services

BACKGROUND

- CHD is the most common major birth defect
- United States Data: 40,000 CHD births /year **
- Majority now survive to adulthood
- 25% of CHD deemed “critical” *
- 4.2% of all neonatal deaths are due to a CHD**
- Nearly half (48%) of the deaths due to CHDs occur during infancy***

*Oster, et al Temporal Trends in Survival in CHD. Pediatrics 2013 May;131(5):e1502-8. doi: 10.1542/peds.2012-3435. Epub 2013 Apr 22.

**US CDC Report September 24, 2010 / 59(37);1208-1211

*** Gilboa et al. Mortality Resulting from CHD in the US '99-'06. Circulation. 2010;122:2254-2263

BACKGROUND

- Practices and surgical results vary among institutions - even within the US
- We now have the ability to recognize “hi-risk” fetuses
- Palliative Care is a relatively young subspecialty – Fetal Palliative care even more so.
- Resources are not infinite
- Just because we can does not mean we should...is intervening always the “right” thing to do?
- “Standard of Care” as a *concept* is evolving, in resource-sufficient and resource-constrained environments alike

Prenatal Diagnosis: Fetal Echocardiography

- Usually performed in the 2nd trimester
- CHD detected in up to ~45% of screened pregnancies*
- Most serious defects easier to detect (Single V ~ 70%)**
- Screening not fool-proof:
 - False positive
 - False-negative
 - Possible progression in-utero
- A thoroughly organized screening program essential to achieve higher detection rates*

*Van Velzen et al. Systematic review and meta-analysis of the performance of second-trimester screening for prenatal detection of congenital heart defects. Int J Gynaecol Obstet. 2018 Feb;140(2):137-14

**<http://pediatrics.aappublications.org/content/pediatrics/early/2015/07/21/peds.2014-3783.full.pdf>

Factors affecting Prenatal Counseling:

- Certainty of diagnosis
- Gestational age
- Presence of Extra-cardiac anomalies
- Presence of Lethal syndromes
- Timing of intervention
 - Fetal life
 - Neonatal period
 - Perhaps later
 - Maybe never!

Our Current Practice at TCH

Obstetricians perform screening ultrasounds



High Risk pregnancies are referred to MFM



Fetuses with suspected cardiac pathology are referred to Fetal Cardiology



Subspecialty referrals made as needed



Weekly multi-disciplinary conferences

Consensus management... and *birth plan*

METHODS

We queried the TCH Fetal Cardiology (FC) and Perinatal Pediatric Advanced Care Team (PPACT) Databases

Abstracted information :

- Number of comprehensive fetal echocardiograms
- Subset of pregnancies at high risk for fetal or neonatal demise
- Number of PPACT referrals
- Chart review of individual cases

RESULTS

- 6314 women were referred to Fetal Cardiology for comprehensive fetal echocardiograms between January of 2015 and 2018
- 14 (0.2%) deemed at high risk for fetal or neonatal demise
- All 14 cases were referred to PFACT
- Targeted consults performed
- Perinatal management plans formulated

OUTCOMES

- 3 intra-uterine fetal deaths
- 2 were still-born
- 7 died within days after delivery
- 2 who were discharged home on hospice care died at home.

100% prediction of fetal or neonatal demise

Which Fetal Diagnoses carry High Risk?

- Seven (50%) had lethal genetic syndromes or co-morbidities :
 - Trisomy 13 (1) or trisomy 18 (2)
 - Congenital High Airway Obstruction (CHAOS – 1)
 - VACTERL with bilateral renal agenesis (1)
 - VACTERL with mitral atresia and severe TR (1)
 - Congenital diaphragmatic hernia with single ventricle (1)

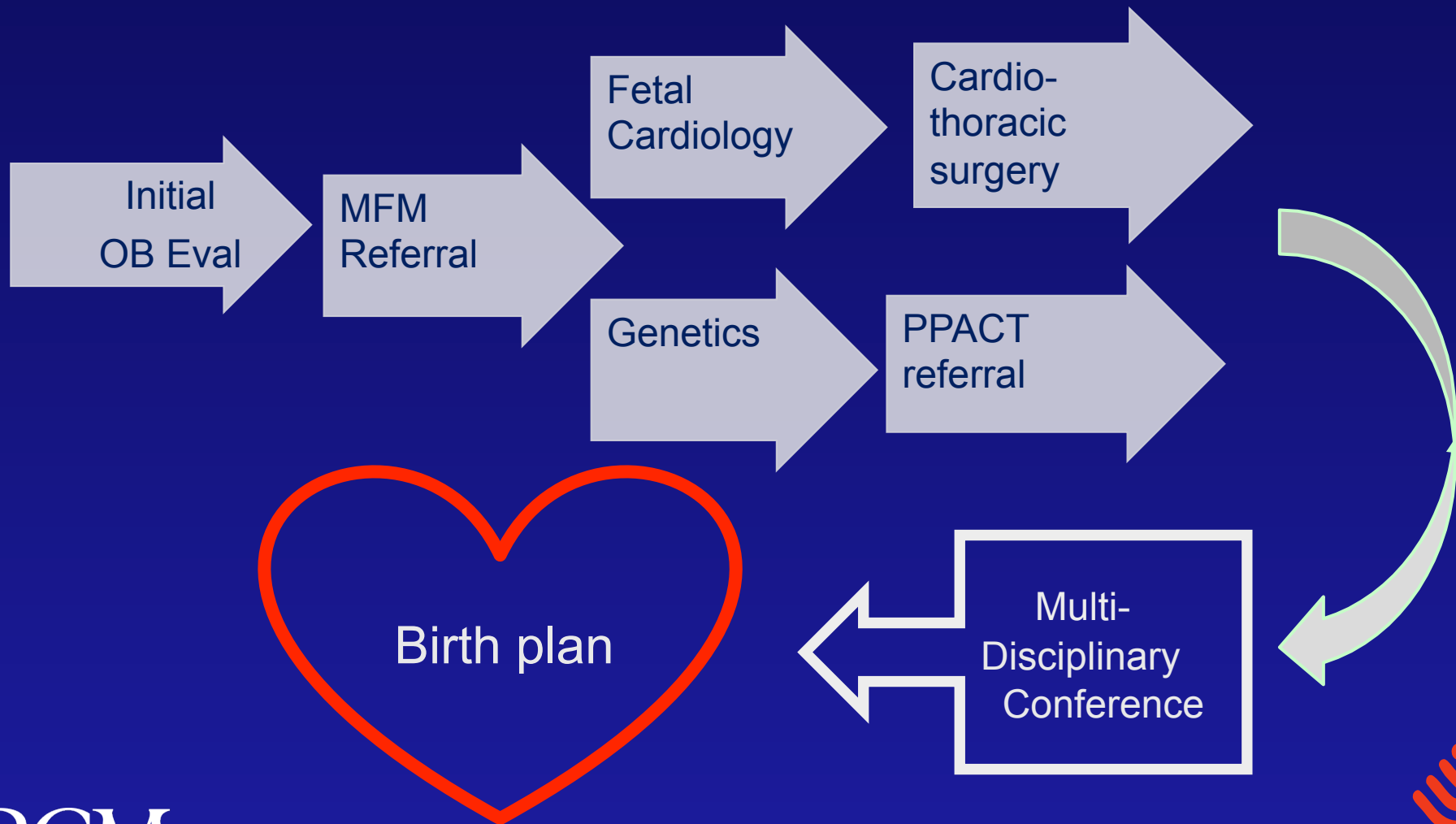
Highest Risk Fetal Cardiac Diagnoses :

- Hypoplastic left heart syndrome with a restrictive atrial septum (2)
- Single ventricle with heart block & hydrops (1)
- Heterotaxy / single ventricle with obstructed anomalously draining pulmonary veins (1)
- Mitral atresia with severe tricuspid regurgitation (2)
- Unbalanced AVSD with severe AV regurgitation (1)

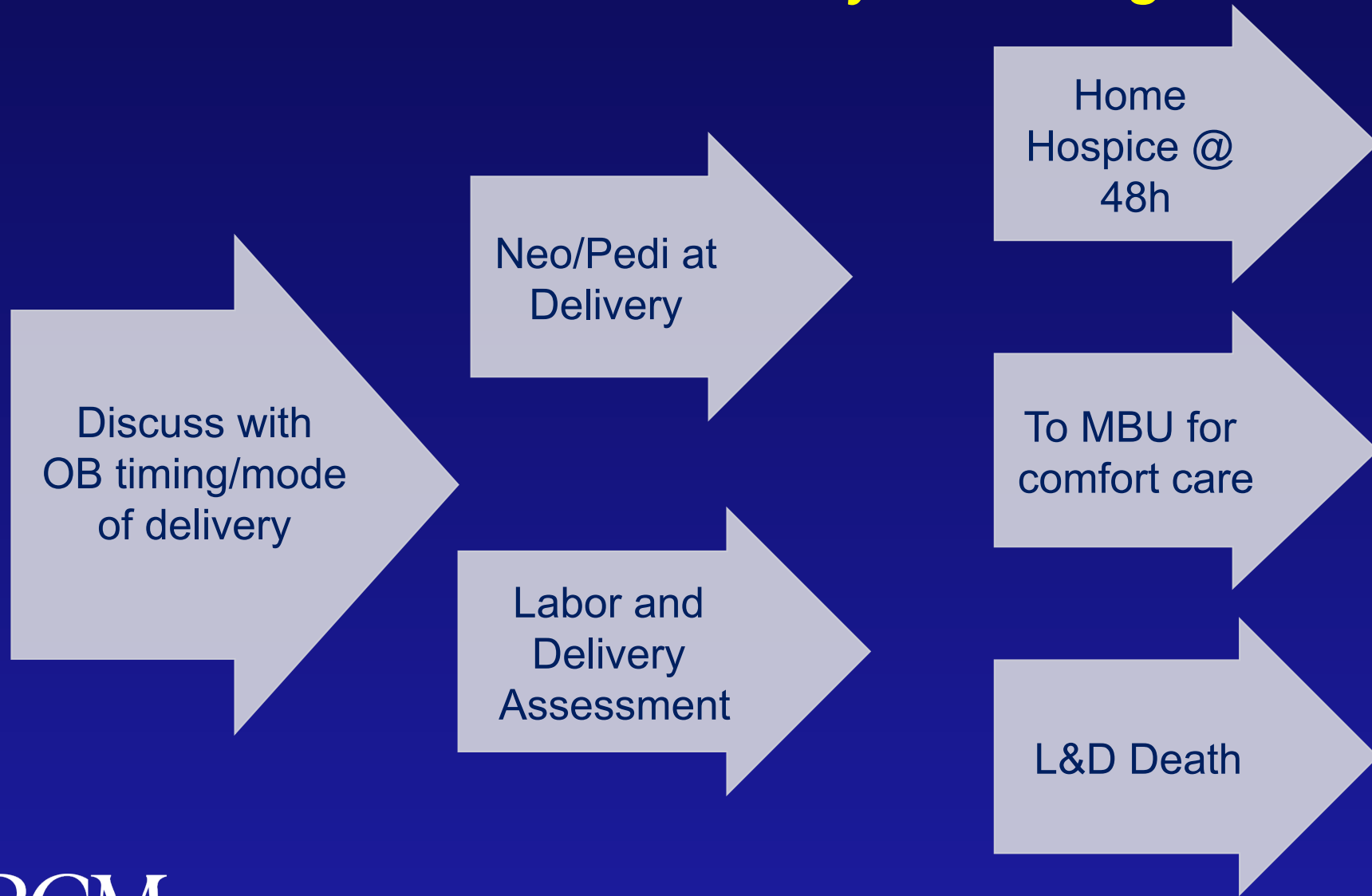
CONCLUSIONS

- Fetal echocardiography reliably predicts severity of lesions, particularly in the third trimester
- The most severe “lethal” lesions account for ~ 0.2% of referrals
- Early referral of these very high-risk pregnancies is recommended, particularly those that would benefit from a *primarily* palliative care approach upon live delivery...
- Seamless birth plans and bereavement care are needed for the best possible outcomes for infants, families, and their medical teams.

Is this the "ideal" practice???



Consensus Delivery Planning



What is the “ideal” practice?

- Mindful of available resources
- Considers local cultural and religious beliefs
- Respectful
- Dignified
- Humane

Thank you!

