Fetal Cardiac Intervention for Hypopatic Left Heart Syndrome

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1. At 20 weeks, on fetal ultrasound you identify an abnormal cardiac structure. Which of the following would exclude the fetus from consideration for "in utero" aortic valve dilation?
   A. retrograde flow in the aortic arch
   B. left ventricle length 50% of right ventricle length
   C. left to right flow across the foramen ovale
   D. sex chromosome aneuploidy

Correct Answer: B

Rationale:
   b) In the subset of fetuses with HLHS, those who originate with a stenotic aortic valve are the best candidates for "in utero" valve dilation. This generally leads to an initially dilated left ventricle which then progress to echogenic myocardium and ventricle hypoplasia later in the third trimester. This is different from the group of newborns with HLHS who have mitral and aortic atresia and a nondeveloped left ventricle from early in pregnancy.
   Other options
   a) Retrograde flow in the aortic arch is a requirement for intervention to assure that fetuses are chosen who with high certainty will progress to HLHS. Often antegrade flow in the arch can be achieved following "in utero" valve dilation
   c) Left to right flow is also a criteria for inclusion to assure a high likelihood that fetuses are proceeding to HLHS if the aortic valve is not dilated
   d) While the majority of fetuses with evolving HLHS are male, there is a higher chance of either 45,X or 45,X mosaicism. Following patient counseling regarding the anticipated clinical effects of 45,X, this alone does not exclude the patient from "in utero" aortic valve dilation.

2. Compared to those children with a univentricular heart, those undergoing "in utero" aortic valve dilation and achieving a two ventricle circulation, are least likely to encounter
   A. growth restriction at birth
   B. neurodevelopment challenges
   C. lower childhood weight and height
   D. early childhood death

Correct Answer: D

Rationale:
   d) With technical success at "in utero" aortic valve dilation, approximately 50% of newborns will be able to sustain systemic circulation with their salvaged left ventricle. These children are able to avoid the morbidity and mortality of the Norwood three staged procedure that converts the circulation to a right side of the heart. Additionally, right sided physiology does not ideally
sustain systemic circulation leading some of the children following a Norwood conversion with a
higher chance of cardiac failure and eventual transplantation with associated morbidity and
mortality.
Additional options
a) Conversion to a biventricular system following a technically successful aortic valve dilation
does not impact fetal growth
b) Neurodevelopmental outcomes are difficult to assess given the multitude of confounders but
do not appear to be different between those children achieving a biventricular circulation
compared to those who progress to HLHS and a univentricular circulation.
c) Challenges of adequate growth, often with a need for assisted feeding as an infant, occur in
both those children achieving biventricular circulation following a fetal "in utero" dilation and
those progressing to univentricular circulation.

References (both questions):
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Fetal imaging of Twins
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1. What is the most common intracranial abnormality in TTTS?
   A. Germinal matrix hemorrhage
   B. Enlargement of the venous sinuses
   C. Choroid plexus cyst
   D. Ventriculomegaly

Correct Answer: D

Rationale:
D. Ventriculomegaly is the most common intracranial abnormality. Tauri et al published that the most frequent abnormality found in TTTS was ventriculomegaly at 63% in both donor and recipient. AJNR Am J Neuroradiol 33:1121–26 Jun-Jul 2012. Altered Fetal Cerebral and Cerebellar Development in Twin-Twin Transfusion Syndrome

Options A and B represent findings commonly seen in TTTS however they are not the most common abnormality.

Option C is not correct because is not a finding related or associated with TTTS.

2. Enlargement of the venous sinuses is seen in:
   A. Donor twin
   B. Recipient twin
   C. Both twins

Correct Answer: C

Rationale:
Enlargement of the venous sinuses is another common finding in twins with TTTS. While it is more commonly seen in the donor (50%), it is also seen in the recipient (13%). Twin–twin transfusion syndrome: cerebral ischemia is not the only fetal MR imaging finding. Kline-Fath et al. Pediatr Radiol (2007) 37:47–56
1. Differential considerations for asymmetric bulging at the placental-uterine interface in the lower uterine segment in addition to MAP include
   A. Adenomyosis
   B. Myometrial window
   C. Both
   D. Neither

Correct Answer: C

Reference:

2. Functional MRI applied to the placenta so far has focused on
   A. Immune mediation
   B. Endocrine regulation
   C. Waste removal
   D. Nutrient delivery

Correct Answer: D

Reference: