Fetal Heart Society

Q&A for FHS Webinar:

Fetal D-TGA: In Utero Physiology, Postnatal Compromise, and Neurodevelopmental Outcomes

Articles Posted in Chat:

- Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management
- Neurodevelopmental evaluation strategies for children with congenital heart disease aged birth through 5 years: recommendations from the cardiac neurodevelopmental outcome collaborative
- Neurodevelopmental evaluation for school-age children with congenital heart disease: recommendations from the cardiac neurodevelopmental outcome collaborative

Q & A:

	Question	Answer
1	In dTGA, if more blood to pulmonary, is it more blood back to pulmonary veins, and make left atrium bigger than not dTGA? Thanks. (Johan Widjaja, Jakarta)	Absolutely correct. foramen ovale flow goes leftward due to increased LAp. If the foramen is restrictive the atrial septum will bow rightward making the LA big.
2	Would you recommend getting brain MRIs on all DTGA infants preop?	Some centers are routinely performing pre- and post-operative brain MRI in patients with d-TGA. My personal opinion is that this information is helpful and can lead to the receipt of early intervention services at the optimal time point. Sometimes the information on the pre-operative MRI can be helpful for the surgeon as well.
3	Could the first speaker expand slightly on his comments about TGA and later risks of pulmonary hypertension, and whether this has to do with the altered TGA fetal physiology? What makes 1 baby develop PH post-ASO, compared to another who doesn't? Thank you!	There is a small subgroup of patients with D-TGA who, even after ASO, have persistent pulmonary hypertension, which can be severe. I don't believe we know exactly why one patient develops PH and another doesn't. The suspicion is that it has to do with abnormal fetal lung blood flow in-utero.
4	I'm in high school right now but I find this webinar really interesting and think that fetal development in general can be impacted multiple ways. Is there any way that people can volunteer or help out this society?	How exciting to have you on! Feel free to email me. mdonofri@childrensnational.org
5	Thoughts on using outcome measures like General Movements at 12 weeks corrected age in this population?	While specific measures aren't named, the assessment working groups of the CNOC certainly discussed assessment of motor skills as an important piece of the puzzle. If you look at the CITY paper we placed in the chat, there is a pre-discharge neurobehavioral exam recommended, and assessment of motor skills is usually a part of this. We do find motor delays and impairments are quite common in infancy/toddlerhood, and early identification and assessment of these is important.

	Question	Answer
6	Will there be more webinars?	The Fetal Heart Society hosts webinars every 2nd Thursday at 8:30 pm Eastern Time.
7	In our early studies in fetuses with maternal hyperoxygenation, not only did the pulmonary artery PI decrease showing increased blood flow, but the pulmonary venous blood flow also dramatically increased	Thank you for your comment. This has been reported as well in a study by the group out of Poland.
8	Can you expand why did prenatal diagnosis was associated with better neurodevelopmental outcomes? Thanks.	We believe it is because the babies are more hemodynamically stable because they have known neonatal CHD vs. those that get picked up after birth and are potentially at risk for hemodynamic instability before getting diagnosed.
9	How reliable is maternal hyper-oxygenation testing in predicting pulmonary hypertension for TGA postnatally?	Good question. This needs to be studied and is a part of the hyperoxia arm of the FHS prospective TGA study. In our pilot study that included 4 patients with TGA, lack of reactivity did predict post-delivery compromise.
10	Since highly oxygenated blood supplying the ductus may contribute to abnormal pulmonary vasculature because of ductal constriction, as well as to foramen valve closure, wouldn't it be especially important to prevent ACQUIRED ductal constriction in TGA reinforcing preventive measures?	This is a great point. I think the question is referring to whether we would cause harm by inducing ductal constriction with maternal hyperoxygenation. Although this is a theoretical risk no study thus far has demonstrated evidence of ductal constriction with brief administration of maternal hyperoxia. However, studies have been small especially in fetuses with TGA. Hopefully through the FHS TGA study we will learn more about potential risk of increases oxygenation on the ductus arteriosus.
11	How do we get better at assessing heterogeneity in placental health and its association with fetal vascular development? How much do you think that's associated with the pulmonary vascular variability or the cerebral vasculature health and ND?	I think this is an area that needs a lot more research moving forward. The placenta is like the forgotten organ and I think it plays a significant role in maternal and fetal health in the context of CHD. At our site we are using fetal MRI, specifically T2* MRI to learn more about the vascularity of the placenta. Others have also published on both pathology of the placenta and advanced imaging techniques such as MRI and there are clearly abnormalities that likely vary between patients depending on genetic and environmental factors. In a paper from Jack Rychik studying placental pathology, their group specifically found that fetuses with d-TGA had significantly small placentas compared to fetuses without CHD. Rychik et al, 2018, Pediatric Cardiology

The following questions were live-answered during the webinar:

- Question for Dr. Schidlow. We know that it is alveolar hypoxia, rather than pulmonary arterial hypoxemia, that causes PA vasoconstriction. I was under the impression that alveolar hyperoxia is responsible for the majority of pulmonary arterial vasodilation, and that the contribution of elevated pulmonary arterial saturation (hyperoxemia) is minimally responsible for vasodilation. Please expand on the data that supports high pulmonary arterial saturation changes PVR and ductal changes. Thanks!

- For David Schidlow, is it standard at Boston Children's to do maternal hyper oxygenation testing for D-TGA and have the results correlated with needing BAS? Also, what's your Master of Music in?

- Is this injury preventable by performing late preterm delivery - 34-36wks?

- Secondly, do we need to reevaluate the stress of labor (transient low oxygenation to fetus)?

- What is considered «late» or «early» for ASO?

- When you say timing of surgery is associated with poor neurological outcomes, is there a specific “day” by which the surgery helps? We have heard the window of 3-5 days. Wondering if that still holds true for neuro outcomes.

- The guidelines mention that up to 3 to 4 weeks the correction of transposition of the great vessels, how does this parallel and hypoxic circulation affect neurodevelopment?

- What is the optimal timing for planned delivery and surgery following fetal diagnosis?

- Early VS late repair as far as neurodevelopment outcome - what is considered late from that stand point? How about the subset of TGA IVS with significant postnatal persistence of fetal circulation and significant pulmonary hypertension (and likely later repair). Do they have worse neurological outcome or is it unknown?

- Thank you for a fascinating webinar! Are there known delivery room risk factors associated with development of pre-op white matter injury?

- I am a physical therapist so I have a little different perspective and I appreciate this lecture very much. I am curious how centers are deciding to refer families to early intervention (PT/OT/SLP/ teachers SI) due to diagnosis vs. waiting for a motor delay i.e. getting early intervention services when a child is younger? There is also a discussion of motor deficits but are the delays evident in a fetus or neonate? Is there research being done in regard to motor development?

- For Brad- any ICU management tips/strategies when baby transferred from a remote hospital with very low sats, on Prostin, intubated?

- We recently had a fetus with d-TGA IVS with intractable PHN postnatally. The baby died on ECMO. Lung biopsy postmortem showed alveolar capillary dysplasia with pulmonary vein malalignment. I wonder if any centers have had a similar experience.