Cardiology Corner: A Prenatal Diagnosis of Transposition of the Great Arteries

Gil Wernovsky MD, FAAP, FACC; Benjamin Hopkins BS (Discussant)

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I want to discuss a recent case involving a full-term baby with a prenatal diagnosis of transposition of the great arteries with an intact ventricular septum (TGA/IVS). The baby was delivered by spontaneous vaginal delivery at term and was transferred to a level 4 NICU in good condition, weighing 3.3 kg, with APGARs of eight and nine, and was a very vigorous baby with no distress. The pulse oximeter recordings in the right arm and right leg were in the high 70s. A prostaglandin infusion was started at 0.05 mcg/kg/min; umbilical venous and arterial catheters were placed. The baby was electively intubated for a balloon atrial septostomy at the bedside within the first 24 hours of age, which proceeded uneventfully. Oxygen saturations rose to the high 80's, in an FIO_2 of 0.4.

A discussion was then had between the staff neonatologist and the consulting cardiologist about the saturation goals. I am always hesitant to answer that question in a dogmatic fashion, as it may result in overtreatment or undertreatment of the underlying pathophysiology in certain situations. Picking a "goal" saturation may oversimplify the unique anatomy and physiology of the particular baby in front of you, and does not take into account the unknowns of the transitional circulation. The question is better framed in the context of not what the saturations *"should"* be, but if they are not as expected, what contributes to the changes in arterial oxygen saturation in *this particular baby* with *this particular disease*? This is particularly important in understanding the implications of a saturation of 70 versus a saturation of 90. Another essential aspect to remember is that you must always state where in the body saturation is being measured in TGA: is it measured in the right hand proximal to the ductus, or is the saturation measured in the foot distal to the ductus?

Transient elevations in the pulmonary vascular resistance in babies with TGA and an open ductus will result in a post-ductal saturation higher than the preductal saturation: so-called "reverse differential cyanosis." From a management perspective, a small difference is of no clinical importance, and management of TGA in any circumstance should be based on the pre-ductal saturation – the blood that supplies the brain and the coronary arteries.

I need to emphasize that each one of these babies with TGA is different in "how the blood goes around," TGA being less predictable than, for example, truncus arteriosus, total anomalous venous return, or hypoplastic left heart syndrome as inter-circulatory mixing is more variable and in TGA than the other three examples. Secondly, the impact of more routine NICU interventions (volume, supplemental oxygen, mechanical ventilation), as well as the transitional circulation and labile pulmonary vascular resistance, change oxygen delivery considerably more than in babies with other forms of CHD and certainly compared to babies with structurally normal hearts.

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Differential Diagnosis of Unacceptable Hypoxemia in a Baby with TGA:

It is important to remember what contributes to the peripheral arterial oxygen saturation in babies with TGA. **The only way oxygenated blood gets from the pulmonary veins to the ascending aorta is through an atrial septal defect (ASD),** which may be the most crucial part of the discussion of this baby. Therefore, in this patient, the upper body oxygen saturation is primarily related to the amount of left atrial return, which is the total pulmonary blood flow. If the saturation is in the 90 range, there is considerable blood flow from the left to the right atrium (RA). If one is dealing with a saturation of 75 percent, there is less blood flow going from the LA to the RA to the ascending aorta.

There are three reasons why there might be decreased LA return going to the right atrium, with resultant unacceptable hypoxemia:

1. Less pulmonary blood flow due to a small patent ductus arteriosus
2. Less shunting of blood from the LA to the RA due to a small intra-atrial communication
3. Less pulmonary blood flow due to elevated or labile pulmonary vascular resistance

Note that the top 2 of these 3 are "cardiac" causes rather than more common pulmonary vascular causes found in most babies in the NICU.

I have seen situations where the prostaglandin at a usual dose has less effect than expected. I have also seen hypoxemic babies where the prostaglandin is correctly ordered, but the infusion is disconnected at the hub and delivered into the bed. I have seen situations where the vascular access is extravascular, and the baby is not receiving prostaglandin. If you are caring for a baby whose circulation is "PGE dependent," the very first thing to check is that the PGE is being delivered in the right place and at the correct dose.

The second thing to assess is if the ASD is adequate in size. If the PDA and atrial septal defect are acceptable, then a baby's oxygen saturation is going to be, in general, 75 to 85 percent in the upper body. If a baby is saturating in the very high range for TGA, say, 88 to 92, that tells you that there is significant pulmonary blood flow, which gives a fair amount of pulmonary venous return and, therefore, LA to RA shunting. However, one must be mindful in a patient with a PDA that if saturations are high (high pulmonary blood flow), frequent assessment of adequate systemic blood flow is very important. As in babies with a structurally normal heart, high flow from the aorta to the pulmonary artery through a PDA puts systemic flow at risk, particularly to the coronaries, brain, and gut.

Back to this Case:

The challenge for the team in this particular baby was on the first post-operative night when the saturations in the right arm were very labile - between 78 to 92. The team correctly diagnosed labile pulmonary vascular resistance physiology and started up to 80 percent supplemental oxygen. The saturations then remained in the low 90 range. About 6 hours later, saturations fell into the mid-80s and inhaled nitric oxide was instituted. The baby became tachycardic and hypotensive and developed an elevated serum lactate. Volume infusions were given, and an epinephrine infusion was begun.

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One of the things I tell my trainees frequently is, "Don't fall in love with your first diagnosis." The first diagnosis, correctly, was labile pulmonary vascular resistance. However, this then resolved and resulted in quite a bit of pulmonary blood flow and intra-atrial shunting with saturations in the 90s. Nonetheless, an additional vasodilator was added to the oxygen, and a significant "systemic steal" occurred into the pulmonary arteries, with resultant low systemic blood flow. The presence of an open ductus and pulmonary vasodilators may have deleterious effects on systemic blood flow. Another important point for all of us in the ICU: if a baby is not responding to the therapies as expected, check your underlying assumptions.

The final point I would like to emphasize is the difference between oxygen saturation in the blood and oxygen delivery to the tissues. Higher oxygen saturations may not be "better" oxygen saturations (although I frequently hear colleagues refer to them as "better") if the systemic blood flow delivering that oxygen is compromised.

In summary, this baby had TGA and an excellent septostomy with an initially labile transitional circulation. The oxygen saturations in the 70s were considered problematic and required treatment. In retrospect, the appropriate management after the septostomy became problematic over the ensuing hours when the pulmonary vascular resistance fell and remained low. Fortunately for the baby, with the removal of the pulmonary vasodilators (supplemental oxygen and nitric oxide), there was a prompt improvement in the blood pressure, systemic oxygen delivery, and the lactate normalized. The baby was then scheduled for an arterial switch, which went well and was uneventful.

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1. An ASD is the only reliable way to get oxygenated blood from the pulmonary veins to the aorta.
2. Instead of asking what the saturation "should" be, the bedside staff should understand all the factors that determine the oxygen saturation and why they might change over time.
3. The combination of a PDA and any type of pulmonary vasodilator – including supplemental oxygen - must be monitored exceptionally cautiously. (This is important for babies with structurally normal hearts as well)
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