Cardiac Corner: Critical Physiological Principles when Caring for Babies with Congenital Heart Diseases

Gil Wernovsky, MD; Benjamin Hopkins, OMSIV (Discussant)

"In this month's edition of Cardiac Corner, I would like to discuss some critical physiological principles necessary for all those caring for babies with congenital heart disease."

In this month's edition of Cardiac Corner, I would like to discuss some critical physiological principles necessary for all those caring for babies with congenital heart disease.

There are three broad concepts which determine chamber and great artery pressures, as well as direction of shunting.

- A hole of any significant size equalizes the pressure on both sides of the hole.
    - "Holes" equalize pressure, but do not determine the direction of shunting

1. Blood rolls "downhill".
    - The differences in the vascular resistance determine the direction of shunting
2. Blue is better than gray.
    - A "low" oxygen saturation with normal cardiac output typically results in improved oxygen delivery than a "normal" oxygen saturation with low systemic blood flow

Let me get into this distinction in more detail. It is not uncommon at the bedside to confuse the crucial distinctions between pressure and resistance. When discussing "holes" such as atrial septal defects, ventricular septal defects, patent ductus arteriosus, and AP window, etc., it is essential to remember that *the pressures are equal on either side of the hole*, particularly at the ventricular and great vessel levels. Therefore, it is also vital to understand the strict definition of pulmonary hypertension: a mean pressure in the pulmonary artery greater than 25 mmHg. Thus, in all patients with a large VSD and with a large patent ductus arteriosus, the pulmonary artery pressure is at the systemic level. Thus, there is "pulmonary hypertension." I will get into this in more detail below.

"When discussing "holes" such as atrial septal defects, ventricular septal defects, patent ductus arteriosus, and AP window, etc., it is essential to remember that the pressures are equal on either side of the hole, particularly at the ventricular and great vessel levels."

The second rule, blood rolls downhill, involves resistance, not pressure. For example, in a baby with a ventricular septal defect, blood will shunt, in most situations, from the left ventricle to the low-resistance pulmonary circuit via the right ventricle. This results in a left to right shunt, pulmonary congestion, and no hypoxemia. If pulmonary vascular resistance is high, or there is an obstruction to pulmonary blood flow, as in Tetralogy of Fallot, blood may go from the right ventricle to the left ventricle, where there is less resistance to flow.

Number three, "blue is better than gray," is the physiologic principle most frequently quoted when discussing complex physiology with my NICU colleagues. By that, we mean that the delivery of oxygen, is more important than the oxygen saturation via pulse oximetry (which, of course, is the percent of hemoglobin, which is bound to oxygen). Indeed, if cardiac output is normal and carrying capacity (hemoglobin) is normal, oxygen saturations in the 60s and 70s, even if sustained, will not result in tissue ischemia, metabolic acidosis, or, importantly, neurologic injury. It is beyond the scope of this article to discuss all of the details of every congenital heart problem. Still, in general, ***not all oxygen saturations that are "higher" are "better."***

The next concept that I'd like to discuss is "shunting." This, by convention in most NICUs, refers to shunting in only one direction, right to left, resulting in hypoxemia, and may be labeled "PPHN". This can easily be determined by pulse oximetry. However, the degree of *left-to-right shunting* cannot be quantified at the bedside but may result in significant clinical illness.

"The next concept that I'd like to discuss is "shunting." This, by convention in most NICUs, refers to shunting in only one direction, right to left, resulting in hypoxemia, and may be labeled "PPHN". This can easily be determined by pulse oximetry. However, the degree of left-to-right shunting cannot be quantitated at the bedside, but may result in significant clinical illness."

I think of "shunting" associated with hypoxemia in two broad categories. The most common scenario in the NICU is *interpulmonary shunting*, where the blood returning from the pulmonary veins is not fully saturated; this is due to lung disease, pneumothorax, pleural effusion, atelectasis, etc. *Intracardiac shunting*, however, results in systemic hypoxemia due to systemic venous return bypassing the pulmonary circulation through an intracardiac or great vessel connection. So, in a hypoxemic newborn with congenital heart disease, it is important to distinguish systemic hypoxemia due to an intrapulmonary shunt, intracardiac shunt, or both.

Finally, "pulmonary hypertension" is a frequently misused term, and I wonder if we will ever get it out of our lexicon. In my world as a congenital cardiologist, pulmonary hypertension needs to be divided into two categories: pulmonary hypertension due to elevated pulmonary vascular resistance (such as seen in PPHN, diaphragmatic hernia, and meconium aspiration), and pulmonary hypertension due to the connection of the ventricles or the great vessels by "holes" (Rule #1), and differences in resistance (Rule #2). For example, echo reports may report "elevated right ventricular and pulmonary artery pressure," which may be assumed by the bedside team that the pressure is elevated due to elevated resistance ("PPHN"), with institution of pulmonary vasodilation. However, it may also be due to Intracardiac or great vessel communications - a very important distinction for management.

As a parting comment, systemic hypoxemia *without* alveolar hypoxia does NOT cause an elevated pulmonary vascular resistance or “worse PPHN” – otherwise, all babies with intracardiac shunts from congenital heart disease would have elevated pulmonary vascular resistance! It is *alveolar hypoxia* which causes elevations in pulmonary vascular resistance, sometimes severe, and should be treated with usual ventilatory maneuvers, inhaled nitric oxide, ECMO, etc. If a baby has hypoxemia with no lung disease, increasing oxygen, non-invasive or invasive mechanical ventilation is likely to do more harm than good.

Disclosure: *The authors have no conflicts of interests to disclose.*

NT

Corresponding Author

Gil Wernovsky MD, FAAP, FACC
Senior Consultant in Cardiac Critical Care and Pediatric Cardiology
Children’s National Hospital
Professor of Pediatrics
George Washington University School of Medicine and Health Sciences
Washington, DC
https://childrensnational.org/visit/find-a-provider/gil-wernovsky
Https://linkedin.com/in/gil-wernovsky-md-1096a0
Email: gwernovsky@childrensnational.org

Corresponding Author

Benjamin Hopkins, OMS IV
Western University of Health Sciences
College of Osteopathic Medicine of the Pacific
Email: Benjamin.Hopkins@westernu.edu