Grunting, Shunting, Confronting
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It is 1980, and I am a neonatal fellow working in 3 neonatal intensive care units (NICUs) in Chicago and Evanston, Illinois.

I have had a lot of clinical experience with a wide variety of patients, but one group of infants I was fascinated with were babies who presented with grunting and shunting and had "Persistence of the Fetal Circulation" (PFC) or "Persistent Pulmonary Hypertension of the Newborn" (PPHN).

The curious thing about them was how sick they were with their hypoxemia and how they required your constant presence at their bedsides as they continued to shunt blood right-to-left past the lungs across the patent ductus arteriosus and the foramen ovale (1).

It was during my fellowship that I talked with my mentor, Dr. Thomas Gardner, and Dr. Abby Adams, one of my fellow colleagues, and we began collecting data on these patients.

At that time, Dr. Bill Fox from Children's Hospital of Philadelphia had written a lot about the diagnosis and management of PFC/PPHN (2). The management approach at that time involved respiratory alkalosis utilizing hyperventilation and subsequently metabolic alkalosis (using sodium bicarbonate), both of which would result in pulmonary vasodilation (2,3).

For all of you who cared for these babies during this time, we all had the experience of strengthening our forearm muscles when the patient would "clamp down" and we would spend hours manually hyperventilating them with an anesthesia bag with an FiO2 of 1.0. We also would "boost" their systemic blood pressure with normal saline boluses and dopamine infusions.

Since we did not have true selective pulmonary vasodilators, we used tolazoline (4, 5), and we also tried nitroglycerin, nitroprusside (6), and other vasoactive drugs with some selective pulmonary vasodilatory properties. (7) However, this led to further bedside stress as maintenance of systemic blood pressures then became another problem. This despite the fact that some of our patients' color dramatically improved with systemic vasodilation.

I also talked about this project with Dr. Mike Schreiber, who at that time was one of my fellow residents, and asked him if he would like to work together. After his residency, he was on his way to the University of California, San Francisco, for his NICU/PICU fellowship, so he respectfully declined.

As a result of our efforts, we collected data from a total of 62 infants with primary or secondary PFC/PPHN, and this series was published in American Journal of Diseases of Children in 1984. (8) We also published a paper about predictors of survival (9), as well as air leak with our aggressive approach with hyperventilation. (10)
The management of PPHN has evolved and become more refined, such that survival has improved from 71% overall in our study (8) to around 90% overall (13). The diagnosis is still based on awareness of at-risk groups and specific clinical findings, with confirmation by echocardiography. As Drs. Lakshminrusimha and Keszler noted in their recent NeoReviews article, management involves treatment of the underlying conditions, such as with surfactant administration; basic ventilator management with avoidance of extreme pH, PCO2, PO2 and base excess; attention and support of systemic circulation; selective pulmonary vasodilation; and if necessary, high frequency ventilation or extracorporeal membrane oxygenation(13,14).

Our first paper is now cited with some frequency as a historical reference about "early efforts" with lower baseline survival rates (7).

References


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