Korean Circ J. 2020 Dec;50(12):1124-1126
https://doi.org/10.4070/kcj.2020.0426
pISSN 1738-5520·eISSN 1738-5555

kcj
Korean Circulation Journal

Editorial

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Congenital Heart Disease in Premature Infants Less than 1,500 grams: It Takes a Delicate Touch from Many Multidisciplinary Experts

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▸ See the article "Morbidity and Mortality of Very Low Birth Weight Infants with Congenital Heart Disease" in volume 50 on page 1113.

OPEN ACCESS

Received: Oct 26, 2020
Accepted: Nov 3, 2020

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Conflict of Interest
The author has no financial conflicts of interest.

The contents of the report are the author's own views and do not necessarily reflect the views of the *Korean Circulation Journal*.

Despite remarkable advancements in surgical skills and the development of tailored devices for very low birth weight (VLBW) infants with congenital heart disease (CHD), treating premature infants weighing less than 1.5 kg at birth is still a challenge. These VLBW infants belong to a particularly vulnerable category among premature infants, even aside from their heart disease.

If we focus on intrauterine myocardial development, the premature myocardium's physiology is quite different from that of the full-term infant's myocardium. The premature heart's unique features include a less organized myocardium with smaller myocytes, fewer sarcomeres per unit mass, and higher water content. Hence, a less compliant and less contractile myocardium compromises hemodynamic status during the postnatal transitional phase. Also, problems from extracardiac systems, including bronchopulmonary dysplasia (BPD), and necrotizing enterocolitis (NEC), and sepsis critically affect the morbidity and mortality of premature infants.

Song et al.[1] reported a morbidity and mortality of VLBW infants with CHD compared to babies from birthweight-matched controls through a neonatologist perspective. It was a single-center retrospective 10-year-cohort study (from 2007 to 2016) of prematurely-born neonates with CHD who underwent critical prematurity care in neonatal intensive care unit (NICU). The mean gestational age (GA) of the infants was 31.1 ± 3.2 weeks, the mean birth weight (BW) was 1,126.2 ± 268.3 g, and 50.6% of the infants were small for their gestational ages.

During the last decade, with improved accuracy and feasibility of antenatal ultrasonography, the centralization of mothers with high-risk pregnancies at tertiary centers for planned deliveries accelerated. This trend was also indirectly reflected in the report that showed a gradual decrease in the number of CHD cases, not to mention complex CHD cases, in the second-tier hospitals.[2] Hence, intuitively, a review of the clinical course of VLBW infants with CHD in a single tertiary center providing consistent protocol may give more reliable and comprehensive information than a multicenter study with heterogeneous treatment strategies in every institution.

Recently, data from the Korea Heart Foundation (KHF) published on the follow-up results of 6,599 CHD cases where the patients were surgically operated on and were financially supported by the KHF between January 2000 and December 2014 were compared in terms of outcome to the previous data.[3] Surgically repaired neonatal cases with complex CHD and high Aristotle Basic Complexity (ABC) and Risk Adjustment for Congenital Heart Surgery-1 scores saw a remarkable increase in better outcomes. Although the neonates were not dichotomized in this study as premature or full-term cases, complex CHD lesions were more common in neonates requiring urgent open-heart surgery which poses a technical challenge, and renders early postoperative mortality.

According to Song et al.'s report,[1] pre-op mortality during the wait for an operation was related to prematurity issues other than the CHD itself. Vigilance to these factors might improve the mortality in this VLBW infant group. They also reported a high mortality of VLBW infants who underwent pulmonary arterial (PA) banding. It is well known that neonates' PA banding in critical condition carries subsequent high mortality. The importance of careful monitoring of PA banding's duration and circumference was stressed a long time ago,[4] and the advantage of PA banding as salvage management in CHD patients with body weight < 2.5 kg was recently revisited.[5]

Among the most notorious prognostic factors affecting long-term prognosis on surviving VLBW infants is BPD. Song et al.[1] also reported that long-term exposure to oxygen, mechanical ventilation, and sepsis were the main factors aggravating BPD irrespective of PDA treatment. A delicate stepwise control of adjuvant oxygen therapy was essential as excessive oxygen or prolonged mandatory mechanical ventilation could trigger or aggravate BPD, especially after completing surgical correction for CHD. We should never forget the ambiguous presentation of sepsis in VLBW premature infants, and vigilance regarding premature infants' signs of infection is also essential. High fatal consequences of NEC are worth mentioning here that NEC is observed more frequently in this group irrespective of the feeding pattern. Similar results are highlighted regarding the difference in pathogenesis mechanism between NEC originated in preterm infants and NEC originated in infants with CHD. Enteral feeding was not associated with NEC in ductal-dependent CHD.[6] A key factor in CHD-related NEC was that CHD infants with NEC was significantly younger as to postnatal age at onset. Also, critically affected sites were different, with splenic flexure in the colon, suggesting ischemia in the watershed area.[7]

Recently, the outcome of premature infants with critical CHD including the new modified version of the Risk Adjustment for Congenital Heart Surgery (M-RACHS) category published [8]. Infants born at less than 32weeks comprised 12.8% of the study population. VLBW, persistent pulmonary hypertension of the newborn, BPD, and M-RACHS 5 or more were associated with in-hospital mortality. In addition to those risk factors, single-ventricular physiology and PA banding were more prevalent in the mortality group among infants who underwent cardiac intervention. Premature babies seem to be particularly vulnerable to PA banding with the aforementioned unique characteristics of premature myocardium.

We get a glimpse of the multisystemic factors affecting the premature heart. Based on recent studies,[1][3][8] not to mention surgical or interventional skills, it seems that the mortality of premature infants with CHD depends more on prematurity management. As the age of premature infants under heart surgery is expected to be getting younger and the number of survived premature infants with critical CHD to be getting increased,[9] multidisciplinary

https://doi.org/10.4070/kcj.2020.0426

experts' approach to NICU care seems to be vital for better outcomes for this unique group of patients. In short, caring for VLBW infants with CHD serves to exemplify the adage, "It takes a village to raise a child."

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