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Incidence of bicuspid aortic valve in 16,185 neonates by echocardiographic screening in a single institution: is it really congenial?

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Background: Bicuspid aortic valve (BAV) has been reported to be the most common congenital heart disease with an incidence of 1 to 2% in the general population; however, its incidence in neonates is still unclear because most of the reported incidence data are based on surgical or autopsy cases in adults.

Purpose: To elucidate the true incidence of congenital isolated BAV in neonates by echocardiographic screening in a population-based study.

Methods: We examined a total of 16,185 full-term neonates (male, 48.9%) born in our institution during either of the following two periods: September 1986 to February 2008 and October 2014 to September 2017. Mean gestational age (range) was 39.4 (36 to 42) weeks, and mean birth weight (range) was 3075 (2268 to 4622) g. On the second day after birth, we performed echocardiography and colour-Doppler flow mapping in all subjects. BAV was classified into two types on the basis of morphologic findings in the parasternal short axis view of the aortic valve: BAV with raphe har-

bouring two cusps and commissures, showing a congenital fusion of two underdeveloped cusps, and BAV without raphe, showing a fish mouth appearance in systolic images. BAV associated with other congenital heart diseases such as coarctation of the aorta was excluded from this study.

Results: BAV was identified in 14 neonates (0.09%), an incidence of 0.9 in 1,000 live births. Of the 14 BAV neonates, five had BAV with raphe, whereas nine had BAV without raphe. Of the five BAV neonates with raphe, four had fusion of the right and noncoronary cusps, whereas one had that of the right and left coronary cusps. No aortic regurgitation was detected except trivial one in only one neonate, and no significant valvar stenosis was detected.

Conclusion: The incidence of BAV in neonates was much lower than previously reported incidence data. An acquired fusion of the cusps may develop in later life, and eventually may result in increased incidence of functional BAV.