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## Critical Congenital Heart Disease, Prevalence, and Detection — Alaska, 2007–2018

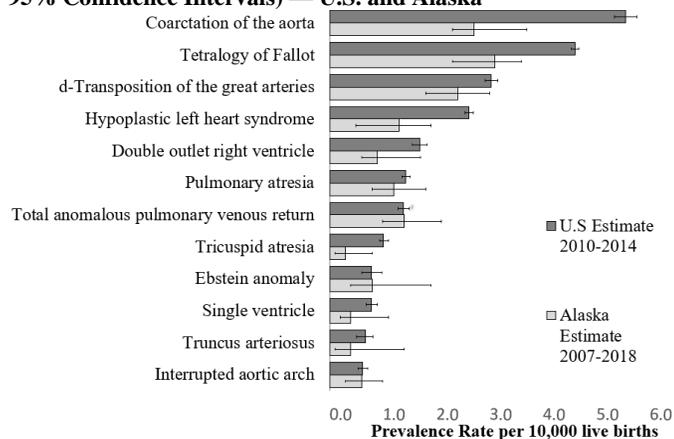
### Introduction

Critical congenital heart defects (CCHDs) are a subgroup of 12 heart defects that require intervention within the first year of life to prevent permanent disability or death.<sup>1</sup> The U.S. Centers for Disease Control and Prevention (CDC) reports that 7,200 infants are born with a CCHD nationally each year.<sup>1</sup> Early detection of CCHDs by prenatal ultrasound, clinical assessment, and pulse oximetry screening performed at 24 hours of life can improve prognosis.<sup>2</sup> This *Bulletin* characterizes the prevalence of CCHDs in Alaska as well as the methods and timing of clinical detection.

### Methods

The Alaska Birth Defects Registry (ABDR) is a passive surveillance system that obtains reports from providers and insurance claims statewide. All birth defects are identified and reported via medical billing codes (ICD-9/10) and are subject to misclassification (false positives). Therefore, a pediatric cardiologist reviewed medical charts on >95% of all reported cases of CCHDs (n=414) for birth years 2007–2018. Confirmed cases of CCHDs (n=221) were used to calculate condition-specific confirmation probabilities, which were applied to all reported cases (including the <5% not reviewed) and used to produce prevalence estimates for all 12 CCHDs. The timing and method of detection of all confirmed cases were summarized. (Note: for more information about the ABDR methods, reports on specific CCHD conditions, and on other important birth defects, visit the [ABDR website](#).)

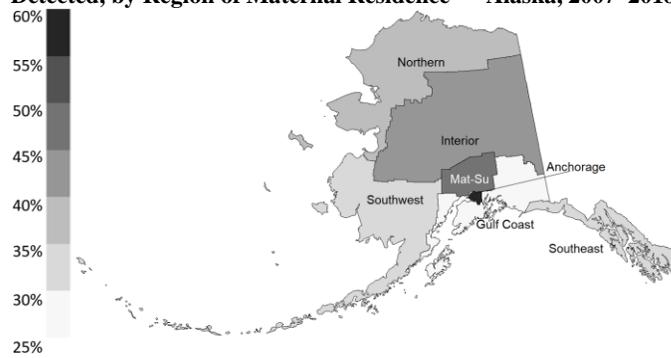
**Figure 1. CCHD Prevalence Estimates per 10,000 Live Births (and 95% Confidence Intervals) — U.S. and Alaska**



### Results

Alaska prevalence estimates for each CCHD were either lower or comparable to national estimates (Figure 1).<sup>3</sup> The prevalence of CCHDs was equivalent across maternal age and race strata and no geographic clustering of CCHDs based on maternal residence was detected (data not shown). Just over half (56%) of confirmed cases of CCHDs in Alaska were reported with at least one additional birth defect (most commonly aneuploidy disorders, limb deformities and reductions, and diaphragmatic hernia). For all confirmed cases of CCHDs, 46.2% were detected prenatally, 24.9% were detected soon after birth during the clinical assessment, 3.6% were detected via pulse oximeter screening at 24-hours post birth, and 23.5% were not detected until after 24-hours after birth (in some cases several months after discharge). Coarctation of the aorta (39%) and Tetralogy of Fallot (25%) accounted for the majority of late CCHD diagnoses. During 2007–2018, the number and proportion of prenatal CCHD detections in Alaska increased over time. Prenatal detection was more frequent in births among residents of more urban regions compared to rural regions (Figure 2).

**Figure 2. Proportion of Confirmed Prenatal CCHD Cases Detected, by Region of Maternal Residence — Alaska, 2007–2018**



### Discussion

Most CCHD prevalence estimates in Alaska are comparable to national estimates, especially for conditions that typically present within the first days of life.<sup>3</sup> However, for CCHDs that may be more difficult to diagnosis early, Alaska's estimates appear to be lower, suggesting a reduced ability to identify these conditions. Detecting a CCHD before a newborn's second day of life is vital for preventing disability or death. While this study found that nearly 24% of CCHDs diagnosed in Alaska during 2007–2018 were detected after 24-hours of life, Alaska has seen improvements in early diagnosis driven by an increase in prenatal ultrasound detection and the 2014 state requirement for pulse oximetry screening. The 4-year average of Alaska's prenatal CCHD detection rate increased by nearly 130% during 2007–2018; this increase follows a national trend.<sup>4</sup> In both Alaska and nationally, prenatal detection of CCHDs is less likely in regions with fewer annual births, resulting in an increase in late detection and less opportunity for delivery at specialized cardiac care centers.<sup>4</sup> These regional differences are most notable for defects that require more advanced sonography imaging, suggesting a training and/or technology gap within the state.

### Recommendations

1. Obstetric and neonatal care providers should ensure they are following The American Academy of Pediatrics CCHD screening protocol, especially in rural areas where prenatal detection of CCHDs tends to occur less frequently compared to urban areas.<sup>5</sup>
2. Prenatal care providers should ensure they are following current American Academy of Family Physicians /American College of Obstetricians and Gynecologists ultrasound screening guidelines and provide sonographers with educational opportunities in both cardiac image acquisition and interpretation.<sup>6</sup>

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