State of Alaska Epidemiology Bulletin

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Bulletin No. 29 November 21, 2016

Prevalence of Congenital Microcephaly — Alaska, 2007–2014

Background

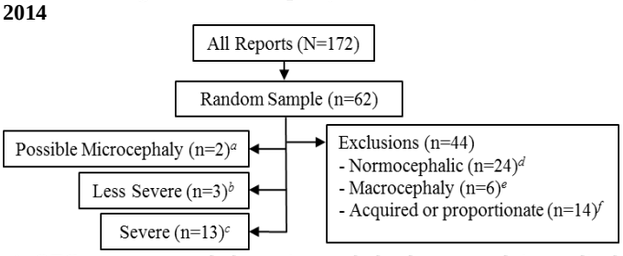
Microcephaly has received increased global attention due to its association with Zika virus infection. According to the National Birth Defects Prevention Network (NBDPN), congenital microcephaly is a clinical diagnosis made prenatally or at birth in a fetus or infant with an occipitofrontal circumference (OFC) that is less than the 3rd percentile for gestational age and sex.[1] Microcephaly can lead to many health problems and early death. During 1996–2011, the estimated prevalence of microcephaly in Alaska, based solely on reported International Classification of Disease (ICD) codes, was 23.6 per 10,000 live births.[2] By comparison, during 2006–2010, the national prevalence ranged from 2–12 per 10,000 live births.[3]

Alaska health care providers are required to report all birth defects using ICD codes to the Alaska Birth Defects Registry (ABDR).[4] Reported ICD codes can misrepresent the prevalence of some birth defects; for example, congenital microcephaly coding is prone to misclassification due to hospital personnel using nonstandard diagnostic criteria and incorrect or imprecise ICD codes. To more accurately estimate of the prevalence of microcephaly in Alaska, we used a novel methodology (based on NBDPN guidelines) that removes the misclassification inherent in calculating prevalence estimates from ICD codes.

Methods

Subsequent to power calculations, we drew a random sample of 36% of microcephaly cases (ICD-9-CM 742.1) reported prior to 2016 and born during 2007–2014 from the ABDR database. We conducted medical records review of the sampled cases and followed NBDPN guidelines for classifying congenital microcephaly based on severity (Figure 1).[1] We estimated the prevalence of congenital microcephaly in Alaska by calculating the case confirmation probability among the sample (confirmed cases/sampled cases) and applied this weight to all reported cases. We estimated the range of variation due to sampling using an alpha of 0.05 with the large-sample binomial approximation.

Figure 1. Congenital Microcephaly Classification, Alaska 2007–2014



a) OFC not measured, but microcephaly documented in medical record, OR OFC measure taken after 6 weeks resulting in microcephaly classification; b) OFC between 3rd and 5th % for age and sex; c) OFC ≤3rd % for age and sex; d) OFC >5th % for age and sex; e) OFC >98th % for age and sex; f) microcephaly developed after birth (acquired), OFC, weight, and length all small for infant's age and sex, but proportional to each other.

Results

Among the 90,481 births that occurred in Alaska during 2007–2014, 172 unique children were reported to ABDR with microcephaly (0.2% of births). Based on reported ICD codes, the prevalence was 19.0 per 10,000 live births (95% CI 16.2–21.8). Among the 62 reported microcephaly cases sampled for confirmation, 13 (21%) met the inclusion criteria for severe congenital microcephaly, and 5 (8%) met the criteria for less severe or possible congenital microcephaly. Based on the case confirmation weights, we estimate the actual prevalence of severe microcephaly in Alaska to be 4.0 per 10,000 births (95% CI 2.2–6.3) or 0.04% of all births. Including less severe and possible microcephaly cases (total congenital microcephaly), the estimated prevalence in Alaska increased to 5.5 per 10,000 births (95% CI 3.5–8.0). During the 8 years of observation, the annual prevalence remained flat (p=0.63 for severe, p=0.60 for total). Regional differences were noted (Figure 2), but no differences were detected by the newborn's sex or maternal age at delivery.

Figure 2. Total Congenital Microcephaly, Alaska 2007–2014

Discussion

During 2007–2014, the prevalence of congenital microcephaly in Alaska ranged from 2–6 and 3–8 for *severe* and *total* cases per 10,000 live births, respectively. These corrected estimates are considerably lower than prior prevalence estimates because they account for coding misclassification. The burden of congenital microcephaly in Alaska (and by region) does not appear to be different than the national burden.[3]

Microcephaly can be detected prenatally by 18–20 weeks gestation, but may not be evident until late in the 2nd trimester. For accurate prenatal diagnosis, serial ultrasounds are advised. The optimal time for OFC measurement is 24–36 hours after birth when head molding subsides, but the measurement is still valid if taken at the 1–2 week well-child check. For congenital microcephaly classification, the OFC must be measured and documented within 6 weeks of birth.[1]

Congenital microcephaly can be caused by genetic problems (e.g., trisomy 13 and 18), maternal malnutrition, teratogens (e.g., maternal phenylketonuria and alcohol use), and *in utero* infections (e.g., cytomegalovirus, toxoplasma, and Zika virus). When microcephaly is suspected, a broad clinical assessment should establish both the cause and severity, as it can occur in combination with other major birth defects.

Recommendations

1. Health care providers should review the Alaska birth defects reporting guidelines (available at: http://dhss.alaska.gov/dph/wcfh/Documents/mchepi/Alaska%20Birth%20Defect%20Registry%20Reporting%20Guide.pdf) and the current NBDPN guidelines for diagnosing and classifying reportable birth defects.[3]
2. Clinicians should work with ABDR to ensure accurate reports and assist with medical case review for conditions like microcephaly that are prone to ICD misclassification.

References

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