

# SENATE BILL REPORT

## SB 6234

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As Passed Senate, February 7, 2024

**Title:** An act relating to screening newborn infants for branched-chain ketoacid dehydrogenase kinase deficiency.

**Brief Description:** Screening newborn infants for branched-chain ketoacid dehydrogenase kinase deficiency.

**Sponsors:** Senators Wilson, L., Hasegawa and Lovick.

**Brief History:**

**Committee Activity:** Health & Long Term Care: 1/25/24, 1/30/24 [DP, w/oRec].

**Floor Activity:** Passed Senate: 2/7/24, 48-0.

**Brief Summary of Bill**

- Directs the State Board of Health to consider whether or not to add the branched-chain ketoacid dehydrogenase kinase deficiency screening to the mandatory newborn screening panel and submit a report the Legislature no later than June 30, 2025.

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### SENATE COMMITTEE ON HEALTH & LONG TERM CARE

**Majority Report:** Do pass.

Signed by Senators Cleveland, Chair; Robinson, Vice Chair; Rivers, Ranking Member; Muzzall, Assistant Ranking Member; Conway, Holy, Padden, Randall and Van De Wege.

**Minority Report:** That it be referred without recommendation.

Signed by Senator Dhingra.

**Staff:** Julie Tran (786-7283)

**Background:** Washington State's Newborn Screenings. Department of Health (DOH)

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*This analysis was prepared by non-partisan legislative staff for the use of legislative members in their deliberations. This analysis is not part of the legislation nor does it constitute a statement of legislative intent.*

must require screening tests of all newborn infants born in any setting. No tests shall be given to any newborn infant whose parents or guardian objects on the grounds of religious tenets and practices.

Annually, DOH performs nearly 12 million tests on more than 172,000 specimens from about 85,000 newborn infants. DOH also provides information and training to parents and health care providers about newborn screenings (NBS). Washington State adds tests to the NBS panel only after a consideration of the following criteria set by the State Board of Health (Board): available technology, diagnostic testing, and treatment available; prevention potential and medical rationale; public health rationale; cost-benefit and cost-effectiveness.

Branched-Chain Ketoacid Dehydrogenase Kinase Deficiency. Branched-chain ketoacid dehydrogenase kinase (BCKDK) deficiency is a genetic condition which causes branched-chain amino acid (BCAA) depletion and the condition is linked to a neurodevelopmental disorder characterized by autism spectrum disorder, intellectual disability, motor impairment, and microcephaly, which is a condition where a baby's head is much smaller than expected.

**Summary of Bill:** The Board must consider whether or not to add the BCKDK deficiency screening to the mandatory NBS panel and submit a report to the Governor and the appropriate committees of the Legislature by no later than June 30, 2025. The report must include an evaluation summary and any findings and recommendations on the addition of BCKDK deficiency to the mandatory NBS panel.

**Appropriation:** None.

**Fiscal Note:** Available.

**Creates Committee/Commission/Task Force that includes Legislative members:** No.

**Effective Date:** Ninety days after adjournment of session in which bill is passed.

**Staff Summary of Public Testimony:** PRO: BCKDK deficiency is linked to autism and to be clear, it is possible to have autism without this deficiency. The BCKDK deficiency is linked to how the brain produces amino acids. Researchers have found that after being provided amino acid supplements, children with autism from BCKDK deficiency saw the symptoms be reversed. If the child is young enough, it can make the autism go away but the difficulty is that an autism diagnosis doesn't happen until children are between the ages 3 to 5, at which point the amino acid treatment isn't as effective. The sooner the children receive treatment, the better the results are. Researchers are advocating for BCKDK deficiency to be tested at birth as a part of the heel prick test, which is when newborns are tested for other rare and treatable disorders. The bill directs the Board to evaluate this condition and decide whether the condition can be added to the screening panel test. Science has come a long

way and with the prevalence of autism rising in recent years, it would be great to have the capability of reversing this trend and the state should at least try and find out if that is possible.

**Persons Testifying:** PRO: Senator Lynda Wilson, Prime Sponsor.

**Persons Signed In To Testify But Not Testifying:** No one.