UNIVERSITY of WASHINGTON
DEPARTMENT OF CHEMISTRY

Michael H. Gelb
Weinstein Endowed Chair in Chemistry

The Honorable Joseline A. Pena-Melnyk
Chair, House Health, and Government Operations Committee
Room 241, House Office Building
Annapolis, MD 21401

Re: HB096 Health - Newborn Screening Program - Krabbe Leukodystrophy

Dear Madam Chairwoman Pena-Melnyk and Distinguished Committee Members:

I want to comment on the precision of newborn screening for Krabbe disease with the most up-to-date information, all of which is published. Newborn screening for Krabbe disease is extremely precise, i.e. essentially no false positives and false negatives. But this requires that the newborn screening lab not only measures the activity of the relevant enzyme in dried blood spots (called GALC enzyme) but for those ~10-20 newborns per year per state who display low GALC, the same blood spot should be used for a second-tier analytics to measure a lipid called psychosine. This can be done by contract with a CLIA-certified lab or in-house in the MD newborn screening lab. It only increases the price of Krabbe newborn screening by a few percent. All babies with severe Krabbe disease show elevated psychosine, and with this two-tier test there are no known false positives or negatives. Furthermore, when psychosine is in the intermediate elevated range the newborn is at high risk for developing a later onset disease and these newborns should be followed in the clinic according to published guidelines written by a team of > 20 experts. This amounts to < 5-10 newborns per year per state so this is a very small number of patients in a follow-up scenario. In summary, newborn screening for extremely precise, in fact more precise than most of the other conditions in the MD newborn screening panel.

Sincerely yours,

Michael H. Gelb
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