77th OREGON LEGISLATIVE ASSEMBLY – 2013 Regular Session **MEASURE: SB 284 A**
STAFF MEASURE SUMMARY **CARRIER: Sen. Boquist**
Senate Committee on Health Care & Human Services

REVENUE: No revenue impact
FISCAL: Minimal fiscal impact, no statement issued

Action:		Do Pass as Amended and Be Printed Engrossed and Rescind the Subsequent Referral to the Committee on Ways and Means
Vote:		5 - 0 - 0
	Yeas:	Knopp, Kruse, Shields, Steiner Hayward, Monnes Anderson
	Nays:	0
	Exc.:	0
Prepared By:		Sandy Thiele-Cirka, Administrator
Meeting Dates:		2/14, 3/11

WHAT THE MEASURE DOES: Directs Oregon Health Authority to adopt rules to conduct two-year study relating to infants who have lysosomal storage disorders. Sunsets March 2, 2015. Declares emergency, effective on passage.

ISSUES DISCUSSED:
- Impact of Krabbe disease
- Newborn screening crucial for early detection
- Current Public Health Laboratory capacity
- Concerns with specificity of disorders in statute rather than administrative rules
- Personal experiences with family and Krabbe disease
- U.S. Department of Health and Human Services, Secretary Advisory Committee on Heritable Disorders in Newborns and Children process
- National efforts on adding Krabbe disease to the screening list
- Routine testing versus diagnostic testing
- Different stages of lysosomal storage disorders

EFFECT OF COMMITTEE AMENDMENT: Replaces measure.

BACKGROUND: Lysosomal storage disorders (LSDs) are inherited disorders caused by a deficiency of specific enzymes that are normally required for the breakdown of certain complex carbohydrates known as glycosaminoglycans (GAGs). If a specific lysosomal enzyme is not present in sufficient quantities, the normal breakdown of GAGs is incomplete or blocked. The cell is then unable to excrete the carbohydrate residues and they accumulate in the lysosomes of the cell. This accumulation disrupts the cell's normal functioning and gives rise to the clinical manifestations of LSDs. LSDs are a group of at least 50 diseases, caused by a genetic deficiency, which causes the accumulation of substances in lysosomes with the cells.

Currently, all newborns in Oregon are screened for 39 diseases at birth through newborn screening. LSD screening is not included. These diseases are not otherwise clinically recognizable at birth, but if untreated can cause irreversible damage both physically and mentally to children. For these diseases, early detection is essential to avoid damaging outcomes.

Senate Bill 284-A directs the Oregon Health Authority (OHA) to conduct a study relating to infants with lysosomal storage disorder and assess the feasibility of requiring newborn screening. OHA is directed to report to the Legislative Assembly on or before March 1, 2015.