WISCONSIN DEPARTMENT OF HEALTH SERVICES
PROPOSED ORDER TO ADOPT PERMANENT AND EMERGENCY RULES

The Wisconsin Department of Health Services (Department) proposes an order to amend DHS 115 (title), s. DHS 115.01, 115.04 (title) and (intro), and create s. DHS 115.04 (8) (cm) and DHS 115.04 (15m), relating to screening of newborns for congenital and metabolic disorders.

FINDING OF EMERGENCY

The Department contracts with the Wisconsin State Laboratory of Hygiene to test most newborns for certain congenital and metabolic disorders that are listed by the Department in s. DHS 115.04. The department proposes to add Carnitine Palmitoyltransferase IA ("CPT IA") and Spinal Muscular Atrophy ("SMA") to the rule.

Preservation of the public health and welfare requires screening newborns for CPT IA and SMA. These disorders present serious health risks in childhood and are unlikely to be detected and prevented in the absence of newborn screening. Interventions are reasonably available at this time, and have been shown, in well-designed studies, to be safe and effective in preventing or ameliorating serious health consequences stemming from a delayed or missed diagnosis of these disorders.

RULE SUMMARY

Statute interpreted
Section 253.13 (1), Stats.

Statutory authority
Sections 253.13 (1) and (4) (b) and 227.11 (2) (a), Stats.

Explanation of agency authority
Under s. 253.13(1), Stats., the Department has the authority to specify the congenital and metabolic disorders for which newborn infants are screened and tested.

Related statute or rule
Section 253.13 (1), Stats.

Plain language analysis
As provided in s. 253.13 (1), Stats., ch. DHS 115 specifies the congenital disorders for which newborns must be screened by means of a blood sample shortly after birth and tested by the Wisconsin State Lab of Hygiene. 2013 Wisconsin Act 135 modified s. 253.13 (1) Stats., relating to infant blood tests so the required screening may be performed by methods in addition to blood testing. Under this emergency and permanent rule order the Department revises ch. DHS 115 to conform the rule language to s. 253.13, Stats. The proposed rule adds Carnitine Palmitoyltransferase IA (CPT IA) deficiency and Spinal Muscular Atrophy (SMA) as conditions for which newborns must be tested.

Summary of, and comparison with, existing or proposed federal regulations
There appears to be no existing or proposed federal regulations that address the activities to be regulated by the emergency rules.

Comparison with rules in adjacent states

Illinois:

Illinois 410 ILCS 240/1.10 (b) 77 Ill. Adm. Code 661.10 *Responsibility for Screening* explains that a Genetic and Metabolic Diseases Advisory Committee will recommend to the Department when an additional disorder should be added to the screening panel. Implementation of the Department's determination is subject to that determination's adoption by rule. This process is similar to Wisconsin's procedure for adding a disorder.

Illinois does screen for CPT1A, but is not currently screening for SMA. However, legislation was passed to screen for SMA, but a testing method is still under development. *See* 410 ILCS 240/3.35.

Iowa:

Iowa Code s. 136A.5A 641—4.3(136A) Iowa newborn screening program (INSP). This program provides comprehensive newborn screening services for hereditary and congenital disorders for the state. 4.3(1) Newborn screening policy. All newborns and infants born in the state of Iowa shall be screened for all congenital and inherited disorders specified by the center and approved by the state board of health.

Iowa does not currently screen for CPT IA or SMA, however, their Congenital and Inherited Disorders Advisory Committee may vote on whether to recommend the addition of SMA to the panel in January, 2019 depending on the availability and outcome of the needs assessment for adding SMA.

Michigan:

Michigan statute, MCLS 333.5430, established a legislatively-mandated advisory committee that is charged with meeting annually. This committee has the authority to add disorders to the NBS panel and approve fee increases associated with adding a disorder.

MCLS, s. 333.5431 (1) (i) refers to CPT IA generally as "other treatable but otherwise disabling conditions as designated by the department." The Michigan Department of Community Health website lists all (55) of the disorders included in their screening panel which includes CPT IA. *See* https://www.michigan.gov/mdhhs/0,5885,7-339-73971_4911_4916-233939--,00.html. CPT-IA is #16 and is found under Fatty Acid Oxidation Disorders.

Michigan has been screening for CPT IA since 2005 but does not currently screen for SMA. However, their legislatively-mandated committee voted to add SMA to Michigan's NBS panel and are working through their approval process.

Minnesota:

Minnesota currently screens for both CPT IA since 2001 and SMA since 2018. The screenings fall under Minnesota's general newborn screening statute (MN 144.125). The process for the newborn screening condition additions consists of an Advisory Committee recommendation to the Commissioner of Health. The Commissioner then has the authority to either accept the recommendation or deny it. If the Commissioner accepts it, then it is added to the panel as of that date and then published in the State Register (see https://mn.gov/admin/assets/SR42_30%20-%20Accessible_tcm36-324655.pdf?sourcePage=%2fadmin%2fbookstore%2fregister.jsp%3fid%3d36-324656).

CPT1A isn't listed since it is not a core condition, but it would have been the same date as all of the other MS/MS conditions (2001). See https://www.health.state.mn.us/people/newbornscreening/program/newbornscreeningpanel.html.

Summary of factual data and analytical methodologies

The Department's Advisory Committee on Newborn Screening (Committee) recommended to the Department, and the Department concurred with the recommendation to add CPT IA and SMA to the list of congenital disorders for which newborns must be screened. Committee members included: Norman Fost, MD (Committee Chair) Ethicist, UW School of Medicine and Public Health, Department of Pediatrics; Mei Baker, MD Co-Director of Newborn Screening Laboratory (WSLH); Jeffrey Britton, MD WI Chapter American Academy of Pediatrics Representative (AAP); Christine Brown Executive Director National PKU Alliance, Parent Representative; Arthur Derse, MD Bioethicist, Medical College of Wisconsin; Ousmane Diallo Office of Health Informatics, Department of Health Services; Kevin Josephson, CGC Genetic Counselor, Gundersen Health System; Steve Leuthner, MD Neonatologist, Medical College of Wisconsin.

Carntine palmitoyltransferase IA (CPT IA) deficiency is a fatty acid oxidation disorder associated with hypoketotic hypoglycemia and liver failure. Children with CPT IA present with hypoglycemia, liver dysfunction and encephalopathy, cholestatic jaundice and hepatomegaly, as well as renal dysfunction manifesting as renal tubular acidosis, when viral illness or prolonged fasting occurs. Prior to the episodes of metabolic crisis, they typically have normal growth and development. Studies have also found evidence for an association between infant mortality associated with infectious disease and homozygosity for CPT IA mutations.

SMA is a neurodegenerative autosomal recessive genetic disease with an estimated incidence of 1 in 10,000 births. It affects the motor neurons in the spinal cord, and involves multiple organs. Cause of death is usually respiratory failure. The Committee determined that CPT IA and SMA met the criteria under s. DHS 115.06 for being added to the list of congenital disorders for which WSLH must test the blood samples of newborns. The former Secretary of the Department of Health Services approved the committee's recommendations, CPT IA on 3/22/2018 and SMA on 6/27/2018.

Analysis and supporting documents used to determine effect on small business

TBD, pending an economic impact analysis.

Effect on small business

The proposed rules are expected to have no effect on small business, pending an economic impact analysis.

Agency contact person

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Statement on quality of agency data

The Department relied on the following information for the rules and analysis: The Department's Advisory Committee on Newborn Screening, the Centers for Disease Control and Prevention, US Secretary of Health and Human Services Department's Advisory Committee on Heritable Disorders in Newborns and Children, and Wisconsin Newborn Screening Program – Condition Nomination Form.

Place where comments are to be submitted and deadline for submission
Comments may be submitted to the agency contact person that is listed above until the deadline given in the upcoming notice of public hearing. The notice of public hearing and deadline for submitting comments will be published in the Wisconsin Administrative Register and to the department's website, at https://www.dhs.wisconsin.gov/rules/permanent.htm. Comments may also be submitted through the Wisconsin Administrative Rules Website, at: https://docs.legis.wisconsin.gov/code/chr/active.

RULE TEXT

SECTION 1. Chapter DHS 115 (title) is amended to read:

(**title)** SCREENING OF NEWBORNS FOR CONGENITAL ~~AND METABOLIC~~ DISORDERS.

SECTION 2. DHS 115.01 is amended to read:

DHS 115.01 Authority and purpose. This chapter is promulgated under the authority of ss. 253.13 (1) and 227.11 (2), Stats., to specify the congenital ~~and metabolic~~ disorders for which each newborn ~~infants are to be~~ is screened and tested.

SECTION 3. DHS 115.04 (title) and (intro.) are amended to read:

DHS 115.04 **Congenital ~~and metabolic~~ disorders**. Pursuant to s. 253.13 (1), Stats., blood samples taken from each newborn shall be tested for all of the following conditions:

SECTION 4. DHS 115.04 (8) (cm) is created to read:

DHS 115.04 **(8) (cm)** Carnitine palmitoyltransferase IA deficiency, ICD-10-CM-E71.318.

SECTION 5. DHS 115.04 (15m) is created to read:

DHS 115.04 **(15m)** Spinal muscular atrophy, ICD-10-CM-G12.9.

SECTION 6. EFFECTIVE DATE. The rules contained in this order shall take effect as emergency rules upon publication in the official state newspaper, as provided in s. 227.24 (1) (c) (intro), Stats.