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# 2011 Risk Updates: FNS Justifications and Implications for WIC Services

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## Underweight

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### Justification

The CDC uses the 2.3rd percentile weight-for-length (for birth to 24 months of age) and the 5th percentile BMI-for-age (for 2-5 years of age), as the cut-offs to define underweight in its Pediatric Nutrition Surveillance System. However, CDC does not have a position regarding the cut-off percentile, which should be used to determine at risk of underweight as a nutrition risk in the WIC Program. At risk of underweight is included in this criterion to reflect the preventive emphasis of the WIC Program.

A review of literature on weight-for-length or stature cut-off percentiles indicates that: a) many children at or below the 5th percentile for weight are in need of nutritional intervention, and b) those at or below the 10th percentile may be at nutritional risk and in need of preventive nutritional intervention, or at least further evaluation.

Weight-for-length/stature describes body proportionality and is sensitive to acute undernutrition, but can also reflect long-term status. Physical growth delay is used as a proxy for the deleterious effects undernutrition can have on immune function, organ development, hormonal function and brain development.

### Implications for WIC Nutrition Services

Participation in WIC has been associated with improved growth in both weight and height in children. An infant or child determined to be underweight at WIC certification should be monitored at regular

intervals during the certification period, as appropriate. Through client-centered counseling, WIC staff can assist families in making nutritionally balanced food choices to promote adequate weight gain. Also, the foods provided by the WIC Program are scientifically-based and intended to address the supplemental nutritional needs of the Program's target population, and can be tailored to meet the needs of individual participants.

In addition, WIC staff can greatly assist families by providing referrals to medical providers and other services, if available, in their community. Such resources may provide the recommended medical assessments, in order to rule out or confirm medical conditions, and offer treatment when necessary and/or in cases where growth improvement is slow to respond to dietary interventions.

## Overweight

113

### Justification

The rapid rise in the prevalence of obesity in children and adolescents is one of the most important public health issues in the United States today. The National Health and Nutrition Examination Survey (NHANES) from the mid-1960s to the early 2000s document a significant increase in obesity among children from preschool age through adolescence. These trends parallel a concurrent increase in obesity among adults, suggesting that fundamental shifts occurring in dietary and/or physical activity behaviors are having an adverse effect on overall energy balance.

The causes of increased obesity rates in the United States are complex. Both genetic make-up and environmental factors contribute to the obesity risk. Important contributors include a large and growing abundance of calorically dense foods and an increased sedentary lifestyle for all ages. Although obesity tends to run in families, a genetic predisposition does not inevitably result in obesity. Environmental and behavioral factors can influence the development of obesity in genetically at-risk people.

BMI is a measure of body weight adjusted for height. While not a direct measure of body fatness, BMI is a useful screening tool to assess adiposity. Children >2 years of age, with a BMI-for-age >85th and <95th percentile are considered overweight and those at or above the 95th percentile, obese. Research on BMI and body fatness shows that the majority of children with BMI-for-age at or above the 95th percentile have high adiposity and less than one-half of the children in the 85th to <95th percentiles have high adiposity. Although an imperfect tool, elevated BMI among children most often indicates increased risk for future adverse health outcomes

and/or development of diseases. BMI should serve as the initial screen and as the starting point for classification of health risks.

Use of the 95th percentile to define obesity identifies those children with a greater likelihood of being obese as adolescents and adults, with increased risk of obesity-related disease and mortality. It is recommended that an obese child (95th percentile) undergo a medical assessment and careful evaluation to identify any underlying health risks or secondary complications. Obesity can result from excessive energy intake, decreased energy expenditure, or a medical condition that impairs the regulation of energy metabolism. In addition, obesity in early childhood may signify problematic feeding practices or evolving family behaviors that, if continued, may contribute to health risks in adulthood related to diet and inactivity.

### **Implications for WIC Nutrition Services**

The WIC Program plays an important role in public health efforts to reduce the prevalence of obesity by actively identifying and enrolling young children who may be obese or at risk of overweight/obesity in later childhood or adolescence. When identifying this risk, it is important to communicate with parents/caregivers in a way that is supportive and nonjudgmental, and with a careful choice of words that convey an empathetic attitude and minimize embarrassment or harm to a child's self-esteem. In recognition of the importance of language, the 2007 American Medical Association Expert Committee Report recommends the use of the terms overweight and obese for documentation and risk assessment only and the use of more neutral terms (e.g., weight disproportional to height, excess weight, BMI) when discussing a child's weight with a parent/caregiver .

BMI is calculated and plotted on growth charts at each WIC certification. However, growth charts are meant to be used as a screening tool and comprise only one aspect of the overall growth

assessment. A clinical assessment to determine if a child is at a healthy weight is more complex. Weight classification (derived from the growth chart) should be integrated with the growth pattern, familial obesity, medical risks, and dietary and physical activity habits to determine the child's obesity risk.

The goal in WIC nutrition counseling is to help the child achieve recommended rates of growth and development. WIC staff can frame the discussion to make achieving normal growth a shared goal of the WIC Program and the parent/caregiver and make clear that obesity is a medical condition that can be addressed.

Parents/caregivers of children may need education on recognition of satiety cues and other physiological needs that lead to crying, and ways to comfort a child (holding, reading, rocking) other than by feeding. The foods provided by the WIC Program are scientifically-based and intended to address the supplemental nutritional needs of the Program's target population and can be tailored to meet the needs of individual participants. Emphasis can be placed on promoting food choices of high nutritional quality while avoiding unnecessary or excessive amounts of calorie rich foods and beverages, and reducing inactivity (like decreasing sedentary TV viewing).

Beliefs about what is an attractive or healthy weight, the importance of physical activity, what foods are desirable or appropriate for parents to provide to children, family mealtime routines, and many other lifestyle habits are influenced by different cultures, and should be considered during the nutrition assessment and counseling. The following resources for obesity prevention can be found at:

◆ Fit WIC Materials:

[http://www.nal.usda.gov/wicworks/Sharing\\_Center/gallery/foodfunfamilies.htm](http://www.nal.usda.gov/wicworks/Sharing_Center/gallery/foodfunfamilies.htm)

◆ MyPryramid for Preschoolers:

<http://www.mypyramid.gov/preschoolers/index.html>

In addition, WIC staff can greatly assist families by providing referrals to medical providers and other services, if available, in their community. Such resources may provide the recommended medical assessments, in order to rule out or confirm medical conditions, and offer treatment when necessary and/or in cases where growth improvement is slow to respond to dietary interventions.

## At Risk of Overweight

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### Justification

The rise in the prevalence of overweight and obesity in children and adolescents is one of the most important public health issues in the United States today. The National Health and Nutrition Examination Survey (NHANES) from the mid-1960s to the early 2000s document a significant increase in overweight among children from preschool age through adolescence. These trends parallel a concurrent increase in obesity among adults, suggesting that fundamental shifts in dietary and/or physical activity behaviors are having an adverse effect on overall energy balance.

BMI is a measure of body weight adjusted for height. While not a direct measure of body fatness, BMI is a useful screening tool to assess adiposity. Children > 2 years of age, with a BMI-for-age > 85th and < 95th percentile are considered overweight and those at or above the 95th percentile, obese. Research on BMI and body fatness shows that the majority of children with BMI-for-age at or above the 95th percentile have high adiposity and less than one-half of the children in the 85th to < 95th percentiles have high adiposity. Although an imperfect tool, elevated BMI among children most often indicates increased risk for future adverse health outcomes and/or development of diseases. BMI should serve as the initial screen and as the starting point for classification of health risks.

Increasingly, attention is being focused on the need for comprehensive strategies that focus on preventing overweight/obesity and a sedentary lifestyle for all ages. Scientific evidence suggests that the presence of obesity in a parent greatly increases the risk of overweight in preschoolers, even when no other overt signs of increasing body mass are present. The presence

of parental obesity should lead to greater efforts by nutrition services staff to assist families in establishing or improving healthy behaviors.

## Implications for WIC Nutrition Services

The WIC Program plays an important role in public health efforts to reduce the prevalence of obesity by actively identifying and enrolling infants and children who may be overweight or at risk of overweight in childhood or adolescence. When identifying this risk, it is important to communicate it in a way that is supportive, nonjudgmental, and with a careful choice of words to convey an empathetic attitude and to minimize embarrassment or harm to a child's self-esteem. In recognition of the importance of language, the 2007 American Medical Association expert committee report recommends the use of the terms overweight and obese for documentation and risk assessment only and the use of more neutral terms (e.g., weight disproportional to height, excess weight, BMI) when discussing a child's weight with a parent/caregiver .

BMI is calculated and plotted on growth charts at each WIC certification. However, growth charts are meant to be used as a screening tool and comprise only one aspect of the overall growth assessment. A clinical assessment to determine if a child is at a healthy weight is more complex. Weight classification (derived from the growth chart) should be integrated with the growth pattern, familial obesity, medical risks, and dietary and physical activity habits to determine the child's obesity risk.

The goal in WIC nutrition counseling is to help the child achieve recommended rates of growth and development. WIC staff can frame the discussion to make achieving normal growth a shared goal of the WIC Program and the parent/caregiver. Studies have shown that the early childhood eating environment provides a great

opportunity for preventive intervention. Parents/caregivers of infants and toddlers may need education on recognition of satiety cues and other physiological needs that lead to crying, and ways to comfort a child (holding, reading, rocking) other than by feeding. Young children look upon their parents as role models for eating behaviors. Through client-centered counseling, WIC staff can emphasize the importance of prevention and can assist families in making changes that improve parenting skills that promote healthy eating, and physical activity behaviors and a healthy weight in children. Also, the foods provided by the WIC Program are scientifically-based and intended to address the supplemental nutritional needs of the Program's target population and can be tailored to meet the needs of individual participants.

Beliefs about what is an attractive or healthy weight, the importance of physical activity, what foods are desirable or appropriate for parents to provide to children, family mealtime routines, and many other lifestyle habits are influenced by different cultures, and should be considered during the nutrition assessment and counseling. The following resources for obesity prevention can be found at:

- ◆ Fit WIC Materials:  
[http://www.nal.usda.gov/wicworks/Sharing\\_Center/gallery/foodfunfamilies.htm](http://www.nal.usda.gov/wicworks/Sharing_Center/gallery/foodfunfamilies.htm)
- ◆ MyPryramid for Preschoolers:  
<http://www.mypyramid.gov/preschoolers/index.html>

In addition, WIC staff can greatly assist families by providing referrals to medical providers and other services, if available, in their community. Such resources may provide the recommended medical assessments, in order to rule out or confirm medical conditions, and offer treatment when necessary and/or in cases where growth improvement is slow to respond to dietary interventions.



## High Weight-for-Length

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### Justification

In 2006, WHO released international growth standards for infants and children aged 0-59 months, similar to the 2000 CDC growth references. Since then, the CDC has developed Birth to 24 months growth charts, based on the WHO growth standards, and recommends their use in the United States. For persons 2-20 years, the 2000 CDC growth charts will continue to be used.

The WHO and CDC growth charts are similar in that both describe weight-for-age, length (or stature)-for-age, weight-for-length (or stature) and body mass index (BMI) for age. However, they differ in the approach taken to create the growth charts. The WHO growth charts are growth standards that describe how healthy children grow under optimal environmental and health conditions. The 2000 CDC charts are a growth reference, not a standard, and describe how certain children grew in a particular place and time.

The WHO growth standards for children < 24 months are based on data collected from 1997-2003 in 6 countries (including the U.S.), from children who were born between 37 and 42 weeks gestation, breastfed for at least 12 months, and introduced to complementary food by at least 6 months but not before 4 months. Infants and children of low-income mothers and/or mothers who smoked were not included in the data sample.

The 2000 CDC charts for infants and children < 36 months are based on birth weight (from 1968 to 1980 and from 1985 to 1994) and birth length data (from 1989 to 1994) obtained from U.S. birth

certificates; National Health and Nutrition Examination Survey (NHANES) data; and, measurements from infants who had been breastfed and formula fed (approximately 50% ever breastfed and approximately 33% who were still breastfeeding at 3 months). Very low birth weight infants were not included in the sample population. This was the only exclusion criterion applied to the sample population.

Prior to making its recommendation, CDC convened an Expert Panel with the National Institutes of Health and the American Academy of Pediatrics to review the scientific evidence and discuss the potential use of the WHO growth standards in the U.S. The recommendation to use WHO growth standards for infants and children < 24 months was made on the basis of input from the Expert Panel. In addition, CDC concluded that the WHO growth standards are based on a high quality study and, since breastfeeding is the recommended infant feeding practice, it is appropriate to use the breastfed infant as the standard against which all other infants are compared.

The WHO growth standards use values of 2 standard deviations away from the median to identify children whose growth might be indicative of adverse health conditions. The CDC Birth to 24 months growth charts (based on the WHO growth standards) labels 2 standard deviations above the median as the 97.7th percentile. Thus, an infant or child (< 24 months) is categorized as high weight-for-length when plotted at or above the 97.7th percentile, labeled as the 98th percentile on the CDC Birth to 24 months growth charts. The CDC recommends that all infants and children < 24 months be assessed using the CDC Birth to 24 months growth charts regardless of type of feeding (formula or breastfed). (See Clarification for information about standard deviations and the cut-off used to determine high weight-for-length.)

## Implications for WIC Nutrition Services

The WIC Program plays an important role in public health efforts to reduce the prevalence of obesity by actively identifying and enrolling infants and young children who may be at risk of overweight/obesity in later childhood or adolescence. When identifying this risk, it is important to communicate with parents/caregivers in a way that is supportive and nonjudgmental, and with a careful choice of words that convey an empathetic attitude and minimize embarrassment or harm to a child's self-esteem. In recognition of the importance of language, the 2007 American Medical Association Expert Committee Report recommends the use of more neutral terms such as weight disproportional to height, excess weight, and high weight-for-length when communicating with a parent/caregiver.

Height and weight measurements are plotted on growth charts at each WIC certification. However, growth charts are meant to be used as a screening tool and comprise only one aspect of the overall growth assessment. A clinical assessment to determine if a child is at a healthy weight is more complex. Weight classification (derived from the growth chart) should be integrated with the growth pattern, familial obesity, medical risks, and dietary and physical activity habits to determine the child's obesity risk.

The goal in WIC nutrition counseling is to help the child achieve recommended rates of growth and development. WIC staff can frame the discussion to make achieving normal growth a shared goal of the WIC Program and the parent/caregiver. Studies have shown that the early childhood eating environment provides a great opportunity for preventive intervention. Parents/caregivers of infants and toddlers may need education on recognition of satiety cues and other physiological needs that lead to crying, and ways to comfort a child (holding, reading, rocking) other than by feeding.

Young children look upon their parents as role models for eating behaviors. Through client-centered counseling, WIC staff can emphasize the importance of prevention and can assist families in making changes that improve parenting skills that promote healthy eating, physical activity behaviors and a healthy weight in children. Also, the foods provided by the WIC Program are scientifically-based and intended to address the supplemental nutritional needs of the Program's target population and can be tailored to meet the needs of individual participants.

Beliefs about what is an attractive or healthy weight, the importance of physical activity, what foods are desirable or appropriate for parents to provide to children, family mealtime routines, and many other lifestyle habits are influenced by different cultures, and should be considered during the nutrition assessment and counseling. The following resources for obesity prevention can be found at:

Fit WIC Materials:

- ◆ [http://www.nal.usda.gov/wicworks/Sharing\\_Center/gallery/foodfunfamilies.htm](http://www.nal.usda.gov/wicworks/Sharing_Center/gallery/foodfunfamilies.htm).
- ◆ *MyPyramid for Preschoolers:* <http://www.mypyramid.gov/preschoolers/index.html>.

In addition, WIC staff can greatly assist families by providing referrals to medical providers and other services, if available, in their community. Such resources may provide the recommended medical assessments, in order to rule out or confirm medical conditions, and offer treatment when necessary and/or in cases where growth improvement is slow to respond to dietary interventions.

## Short Stature

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### Justification

The CDC uses the 2.3rd percentile (for birth to 24 months of age) and the 5th percentile (for 2-5 years of age) stature-for-age, as the cut-offs to define short stature in its Pediatric Nutrition Surveillance System. However, CDC does not have a position regarding the cut-off percentile which should be used to determine at risk of short stature as a nutritional risk in the WIC Program. At risk of short stature is included in this criterion to reflect the preventive emphasis of the WIC Program.

Abnormally short stature in infants and children is widely recognized as a response to an inadequate nutrient supply at the cellular level. This indicator can help identify children whose growth is stunted due to prolonged undernutrition or repeated illness. Short stature is related to a lack of total dietary energy and to poor dietary quality that provides inadequate protein, particularly animal protein, and inadequate amounts of micronutrients such as zinc, vitamin A, iron, copper, iodine, calcium, and phosphorus. In these circumstances, maintenance of basic metabolic functions takes precedence, and thus resources are diverted from linear growth.

Demonstrable differences in stature exist among children of different ethnic and racial groups. However, racial and ethnic differences are relatively minor compared with environmental factors. Growth patterns of children of racial groups whose short stature has traditionally been attributed to genetics have been observed to increase in rate and in final height under conditions of improved nutrition.

Short stature may also result from disease conditions such as endocrine disturbances, inborn errors of metabolism, intrinsic bone

diseases, chromosomal defects, fetal alcohol syndrome, and chronic systemic diseases.

## Implications for WIC Nutrition Services

Participation in WIC has been associated with improved growth in both weight and height in children. A more in-depth dietary assessment and/or referral to a health care provider may be necessary to determine if short stature is a result of dietary inadequacy or a disease condition. Also, more frequent follow-up to monitor growth is appropriate for children in these categories. Through client-centered counseling WIC staff can assist families in improving dietary intake to promote healthy growth and development. In addition, the foods provided by the WIC Program are scientifically-based and intended to address the supplemental nutritional needs of the Program's target population, and can be tailored to meet the needs of individual participants.

In addition, WIC staff can greatly assist families by providing referrals to medical providers and other services, if available, in their community. Such resources may provide the recommended medical assessments, in order to rule out or confirm medical conditions, and offer treatment when necessary and/or in cases where growth improvement is slow to respond to dietary interventions.

# Thyroid Disorders

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## Justification

The thyroid gland manufactures three thyroid hormones: thyroxine (T4), triiodothyronine (T3), and calcitonin. The thyroid hormones regulate how the body gets energy from food (metabolism). Iodine is an essential component of the T4 and T3 hormones and must come from the diet. (Note: In nature, iodine does not exist as a free element; rather, it forms compounds such as sodium iodide. For more information see Clarification section.) Iodine is available from various foods, and is present naturally in soil and sea water. A dysfunctional thyroid gland can become enlarged (goiter) as a result of an overproduction of thyroid hormones (hyperthyroidism) or conversely, from insufficient thyroid hormone production (hypothyroidism). Thyroid hormones influence virtually every organ system in the body.

Maternal needs for dietary iodine and thyroid hormone medication (if prescribed) increase during pregnancy as maternal thyroid hormones and iodine are transferred to the fetus along with an increased loss of iodine through the maternal kidneys. Concurrently, the fetus is unable to produce thyroid hormones during the first trimester and is entirely dependent on the maternal supply of thyroid hormones. As a result, maternal production of T4 must increase by at least 50% during pregnancy. If the pregnant woman is receiving thyroid hormone therapy, often a 30% - 50% increase in thyroid hormone medication is also needed.

### Hyperthyroidism

Hyperthyroidism is a condition in which the thyroid gland is overactive, manufacturing too much thyroid hormone (T4 and T3). An excessive consumption of iodine ( $> 1000 \mu\text{g}/\text{d}$ ) may cause fetal and maternal hyperthyroidism. In other circumstances, the thyroid

might develop nodules which secrete excessive amounts of thyroid hormone regardless of iodine status. Enlargement of the thyroid gland (goiter) is a common symptom, as well as weight loss, fatigue, muscle weakness and an irregular heartbeat.

Hyperthyroidism is relatively uncommon in pregnancy. However, when it occurs, uncontrolled hyperthyroidism (especially in the second half of pregnancy) may result in infection, miscarriage, preterm delivery, preeclampsia, or congestive heart failure. Fetal complications may include prematurity, small for gestational age, fetal or neonatal thyrotoxicosis, or death. Postpartum maternal hyperthyroidism is likely in women with prenatal hyperthyroidism.

The primary medical therapy for hyperthyroidism is radioactive iodine therapy which is contraindicated during pregnancy and lactation. If hyperthyroidism occurs during this period, low doses of thiomide (antithyroid drug) are given instead.

### **Hypothyroidism**

Hypothyroidism is a condition in which the thyroid gland does not make enough thyroid hormone. Maternal and fetal hypothyroidism may occur when preconception maternal iodine stores are insufficient and there is inadequate maternal iodine intake in early pregnancy. In this instance, the maternal iodine balance may become negative and may never be restored, even with eventual iodine supplementation.

Mothers with iodine deficiency during the first half of pregnancy may produce offspring with severe, irreversible brain damage. Maternal thyroid deficiency has been associated with neonatal developmental problems which may cause lasting changes in the brain structure and cognitive function.

Uncontrolled hypothyroidism in the second half of pregnancy can cause maternal complications such as anemia, preeclampsia, miscarriage, premature delivery, and postpartum thyroid disease.

Fetal or neonatal complications include prematurity, low birth weight, congenital anomalies, poor neuropsychological development, and stillbirth.

When iodine nutrition status is adequate, autoimmune thyroid disease (AITD) – also called Hashimoto’s thyroiditis - is the most common type of hypothyroidism during pregnancy. Pregnant women with AITD are at increased risk of miscarriage and postpartum thyroid disease (including thyroiditis, hyperthyroidism and hypothyroidism). There is an increased risk of permanent and significant impairment in cognitive function for their infants.

### **Congenital Hyperthyroidism and Hypothyroidism**

Congenital hyperthyroidism is rare in neonates. Transient congenital hyperthyroidism is caused by maternal Graves disease. Thyroid stimulating immunoglobulin passes from the mother to the fetus via the placenta and causes thyrotoxicosis in the fetus and subsequently, the neonate. After the baby is born, improvement is rapid if the condition is treated using antithyroid drugs and the hyperthyroidism will subside within several weeks. Persistent congenital hyperthyroidism is a familial non-autoimmune disease. It is caused by a genetic mutation resulting in an increase in the constitutive activity of the TSH receptor.

Congenital hypothyroidism due to maternal iodine deficiency is a leading cause of preventable mental retardation. Over-treatment of thyroid hormone, during pregnancy, as well as prolonged maternal iodine therapy (more than two weeks of therapy or more than 1000 µg/iodine) can also cause congenital hypothyroidism. The condition is exacerbated by coexisting selenium and vitamin A deficiencies or iron deficiency. Treatment for neonatal hypothyroidism should be started as soon as possible, as every day of delay may result in loss of IQ. Unless treated shortly after birth (within the first 18 days of life), the resulting mental retardation will be irreversible.

### **Postpartum Thyroiditis**

Postpartum thyroiditis, an autoimmune inflammation of the thyroid, occurs within the first year after delivery or sometimes after termination of pregnancy. It can be a transient thyroid dysfunction with a brief thyrotoxic phase followed by hypothyroidism, usually with a spontaneous resolution. Smoking is a significant precipitating factor in the onset of postpartum thyroiditis. Women with a past history of postpartum thyroiditis have a risk of long-term permanent hypothyroidism and recurrence of postpartum thyroiditis in subsequent pregnancies. Tests for this condition consist of radioactive products necessitating a temporary cessation of breastfeeding (usually up to 3 days).

### **Implications for WIC Nutrition Services**

Individuals with thyroid disorders can benefit from WIC foods and WIC nutrition services can reinforce and support the medical and dietary therapy prescribed by the participants' health care provider. The following nutrition education messages may be appropriate depending on the type of thyroid disorder:

- ◆ Encourage iodine sufficiency, unless contraindicated, with an adequate intake of foods high in iodine such as iodized table salt, bread, saltwater fish, kelp, egg yolks (because of iodine supplementation in chicken feed), milk and milk products (because of the treatment of cows with supplemental dietary iodine). It is important to note that the salt used in manufactured foods is not iodized.
- ◆ Advise women to review the iodine content of their prenatal supplement. It is recommended that all prenatal vitamin-mineral supplements for use during pregnancy and lactation contain at least 150 micrograms of iodine a day. Currently, less than 50 percent of prenatal vitamins on the market contain iodine.

- ◆ Promote breastfeeding, as there are no contraindications to breastfeeding and thyroid hormone replacement therapy as long as normal thyroxine levels in the maternal plasma are maintained. Breast milk provides iodine to the infant and is influenced by the dietary intake of the pregnant and lactating mother. Hyperthyroidism can develop for the first time during the postpartum period, but the mother's ability to lactate is not affected. However, if a woman with untreated hypothyroidism breastfeeds, her milk supply may be insufficient. In such instances, replacement thyroid hormone therapy is necessary to help increase milk production.
  
- ◆ Weight management - hyperthyroidism: The elevated plasma levels of thyroid hormones may cause increased energy expenditure and weight loss along with increased appetite. Following medical treatment, individuals with hyperthyroidism usually regain their typical body weight with a concurrent decrease in appetite. Therefore, the monitoring of weight status and dietary adequacy are recommended.
  
- ◆ Weight management – hypothyroidism: Many individuals with hypothyroidism experience an increase in weight due to both a decrease in basal metabolic rate and an excessive accumulation of water and salt. Most of the weight gained is due to the excess water and salt retention. After medical treatment, a small amount of weight may be lost, usually less than 10% of body weight. Once hypothyroidism has been treated and thyroid hormones are within normal levels, it is less likely that the weight gain is solely due to the thyroid. If an overweight condition persists, weight control therapy may be necessary.
  
- ◆ Recommend the cautionary use of soy formula and the avoidance of foods or supplements rich in soy, fiber, or iron when therapeutic thyroid medications are prescribed, since soy, iron, calcium, fiber and phytates may interfere with the absorption of oral thyroid hormone therapy.

- ◆ Discourage smoking as the compound thiocyanate found in tobacco smoke inhibits iodine transport.

# Inborn Errors of Metabolism

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## Justification

The inheritance of most metabolic disorders is rare. IEM disorders may manifest at any stage of life, from infancy to adulthood. Early identification of IEM correlates with significant reduction in morbidity, mortality, and associated disabilities for those affected.

All States screen newborns for IEM, although the type and number of IEM screened for may vary from State to State. Typically, infants are screened for amino acid disorders, urea cycle disorders, organic acid disorders, and fatty acid oxidation defects. A few States are working toward including lysosomal storage diseases and peroxisomal disorders among their newborn screening panels.

In most states, treatment of an IEM is referred to a specialized metabolic treatment facility. Please see Clarification for contact information for treatment facilities. IEM treatment is based on symptomatic therapy which may include the following strategies: substrate restriction; stimulation or stabilization of residual enzyme activity; replacement of deficient products; removal of toxic metabolites or blocking their production; and enzyme replacement therapy. Avoidance of catabolism is essential at all treatment stages.

Nutrition therapy is integral to the treatment of IEM. Nutrition therapy should both correct the metabolic imbalance and ensure adequate energy, protein, and nutrients for normal growth and development among affected individuals. Continual monitoring of nutrient intake, laboratory values, and the individual's growth are needed for evaluation of the adequacy of the prescribed diet. It is important that caregivers of infants and children with IEM ensure that the patient follows the prescribed dietary regimen. The below

embedded links provide the most up-to-date information about the disease state as well as treatment.

### **Amino Acid Metabolism Disorders**

- ◆ [\*Phenylketonuria \(includes clinically significant hyperphenylalaninemia variants\)\*](#)
- ◆ [\*Maple syrup urine disease\*](#)
- ◆ [\*Homocystinuria\*](#)
- ◆ [\*Tyrosinemia\*](#)

Amino Acid Metabolism Disorders are characterized by the inability to metabolize a certain essential amino acid. The build-up of the amino acid that is not metabolized can be toxic. Treatment of amino acid disorders involves restricting one or more essential amino acids to the minimum required for growth and development and supplying the missing product due to the blocked reaction.

### **Carbohydrate Disorders**

- ◆ [\*Galactosemia\*](#)
- ◆ [\*Glycogen storage disease type I\*](#)
- ◆ [\*Glycogen storage disease type II \(See also Pompe disease\)\*](#)
- ◆ [\*Glycogen storage disease type III\*](#)
- ◆ [\*Glycogen storage disease type IV \(Andersen Disease\)\*](#)
- ◆ [\*Glycogen storage disease type V\*](#)
- ◆ [\*Glycogen storage disease type VI\*](#)
- ◆ [\*Hereditary Fructose Intolerance \(Fructose 1-phosphate aldolase deficiency, Fructose 1, 6, biphosphatase deficiency, fructose kinase deficiency\)\*](#)

This group of disorders includes an enzyme deficiency or its cofactor that affects the catabolism or anabolism of carbohydrate. Carbohydrate disorders are complex and affect neurological, physical, and nutritional status.

### **Fatty Acid Oxidation Defects**

- ◆ [\*Medium-chain acyl-CoA dehydrogenase deficiency\*](#)
- ◆ [\*Long-chain 3-hydroxyacyl-CoA dehydrogenase deficiency\*](#)

- ◆ [\*Trifunctional protein deficiency type 1 \(LCHAD deficiency\)\*](#)
- ◆ [\*Trifunctional protein deficiency type 2 \(mitochondrial trifunctional protein deficiency\)\*](#)
- ◆ [\*Carnitine uptake defect \(primary carnitine deficiency\)\*](#)
- ◆ [\*Very long-chain acyl-CoA dehydrogenase deficiency\*](#)

Fatty acid oxidation defects include any enzyme defect in the process of mitochondrial fatty acid oxidation (FAO) system. The biochemical characteristic of all FAO defects is abnormal low ketone production as a result of the increased energy demands. This results in fasting hypoglycemia with severe acidosis secondary to the abnormal accumulation of intermediate metabolites of FAO, which can result in death.

### **Organic Acid Disorders (AKA organic aciduria or organic acidemia)**

- ◆ [\*Isovaleric acidemia\*](#)
- ◆ [\*3-Methylcrotonyl-CoA carboxylase deficiency\*](#)
- ◆ [\*Glutaric acidemia type I\*](#)
- ◆ [\*Glutaric acidemia type II\*](#)
- ◆ [\*3-hydroxy-3-methylglutaryl-coenzyme A lyase deficiency\*](#)
- ◆ [\*Multiple carboxylase deficiency \(Biotinidase deficiency, Holocarboxylase synthetase deficiency\)\*](#)
- ◆ [\*Methylmalonic acidemia\*](#)
- ◆ [\*Propionic acidemia\*](#)
- ◆ [\*Beta-ketothiolase deficiency\*](#)

Organic Acid Disorders are characterized by the excretion of non-amino organic acids in the urine. Most of the disorders are caused by a deficient enzyme involving the catabolism of specific amino acid(s). As a result, the non-metabolized substance accumulates due to the blockage of the specific metabolic pathway, which is toxic to certain organs and may also cause damage to the brain.

### **Lysosomal Storage Diseases**

- ◆ [\*Fabry disease \( \$\alpha\$ -galactosidase A deficiency\)\*](#)
- ◆ [\*Gauchers disease \(glucocerebrosidase deficiency\)\*](#)

- ◆ Pompe disease (glycogen storage disease Type II, or acid  $\alpha$ -glucosidase deficiency)

Lysosomal storage diseases are a group of related conditions characterized by increased storage of undigested large molecule in lysosomes. Lysosome is a cellular organelle responsible for intracellular degradation and recycling of macromolecules. Due to a defect in a specific lysosomal enzyme, the macromolecule that normally would be metabolized is not broken down; instead, it accumulates in the lysosomes. This leads to tissue damage, organ failures and premature death. Common clinical features include bone abnormalities, organomegaly, developmental impairment and central, peripheral nervous system disorders.

### **Mitochondrial Disorders**

- ◆ [\*Leber hereditary optic neuropathy\*](#)
- ◆ [\*Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes \(MELAS\)\*](#)
- ◆ [\*Mitochondrial neurogastrointestinal encephalopathy disease \(MNGIE\)\*](#)
- ◆ [\*Myoclonic epilepsy with ragged-red fibers \(MERRF\)\*](#)
- ◆ [\*Neuropathy, ataxia, and retinitis pigmentosa \(NARP\)\*](#)
- ◆ [\*Pyruvate carboxylase deficiency\*](#)

Mitochondrial Disorders are caused by the dysfunction of the mitochondrial respiratory chain, or electron transport chain (ETC). Mitochondria play an essential role in energy production. The ETC dysfunction increases free radical production, which causes mitochondrial cellular damage, cell death and tissue necrosis and further worsens ETC dysfunction and thus forms a vicious cycle. The disorders can affect almost all organ systems. However, the organs and cells that have the highest energy demand, such as the brain and muscles (skeletal and cardiac) are most affected. The clinical features vary greatly among this group of disorders, but most have multiple organ dysfunctions with severe neuropathy and myopathy.

## Peroxisomal Disorders

- ◆ [\*Zellweger Syndrome Spectrum\*](#)
- ◆ [\*Adrenoleukodystrophy \(x-ALD\)\*](#)

There are two types of peroxisomal disorders: single peroxisomal enzyme deficiencies and peroxisomal biogenesis disorders. These disorders cause severe seizures and psychomotor retardation. Peroxisomes are small organelles found in cytoplasm of all cells. They carry out oxidative reactions which generate hydrogen peroxides. They also contain catalase (peroxidase), which is important in detoxifying ethanol, formic acid and other toxins. Single peroxisomal enzyme deficiencies are diseases with dysfunction of a specific enzyme, such as acyl coenzyme A oxidase deficiency. Peroxisomal biogenesis disorders are caused by multiple peroxisome enzymes such as Zellweger syndrome and neonatal adrenoleukodystrophy.

## Urea Cycle Disorders

- ◆ [\*Citrullinemia\*](#)
- ◆ [\*Argininosuccinic aciduria\*](#)
- ◆ [\*Carbamoyl phosphate synthetase I deficiency\*](#)

Urea Cycle Disorders occur when any defect or total absence of any of the enzymes or the cofactors used in the urea cycle results in the accumulation of ammonia in the blood. The urea cycle converts waste nitrogen into urea and excretes it from the kidneys. Since there are no alternate pathways to clear the ammonia, dysfunction of the urea cycle results in neurologic damages.

## Implications for WIC Nutrition Services

WIC can provide exempt infant formulas and WIC-eligible medical foods, including those specifically formulated for IEM. Most of the dietary regimens for IEM require a combination of medical food (special formula in most cases) and standard infant formula or prescribed conventional foods. For example, participants with IEM related to essential amino acid metabolism (such as PKU, MSUD), who are not developmentally ready for conventional foods; require both medical food without the offending amino acid(s), and human milk or standard infant formula.

It is recommended that WIC nutritionists collaborate with the clinical dietitians at the metabolic treatment facility, where available, to prescribe WIC food packages (Food Package III) according to the therapeutic diet ordered by the metabolic team, monitor the compliance of the restricted diet, and follow up on the growth and developmental status of the participants with IEM.

Note: Infants with classic galactosemia cannot be breastfed due to lactose in human milk.