Jaundice

Jaundice is clinically apparent when there is a yellowish discoloration of the skin, sclera, and mucous membranes and is evident when the total bilirubin level rises above 4-5 mg/dl in infants and 3 mg/dl in older children. It is important to identify the cause of excess bilirubin in order to initiate appropriate treatment. Elevated bilirubin levels should always be fractionated into unconjugated (indirect) or conjugated (direct) bilirubin and classified as pre-hepatic, hepatic, or post-hepatic. Pre-hepatic jaundice arises when the excess levels overwhelm the hepatocyte's ability to conjugate bilirubin. Hepatic jaundice presents when there is failure of bile metabolism or excretion. Post-hepatic jaundice occurs when there is interruption of bile drainage into the biliary system.

Jaundice can occur in all age groups but is more prevalent in neonates and commonly results from the accumulation of unconjugated bilirubin. Neonatal jaundice appears from either increased bilirubin production or decreased secretion. Increased production may be caused from fetal-maternal blood group incompatibilities, hemorrhage, polycythemia, red blood cell abnormalities, or labor induction. Decreased

Youhanna Al-Tawil , MD
Misty Murphy FNP-C

bilirubin excretion may be directly related to breast-feeding, inborn errors of metabolism, hormones, drugs, prematurity, hepatic hypoperfusion, cholestatic syndromes, or biliary tree obstructions. Sepsis, intrauterine infection, and congenital cirrhosis may also be the culprit.

Physiologic jaundice manifests after the first 24 hours of life. This is commonly breastfeeding induced and is associated with suboptimal milk and caloric intake. Jaundice manifesting before the first 24 hours is always considered pathologic until proven otherwise. Breast milk jaundice manifests after the first 4-7 days after birth and there is a peak in levels around 14 days. In the clinic setting, identifying BMJ is important. The clinician needs to educate the family on symptoms of dehydration secondary to inadequate breastmilk intake and poor feeding. When jaundice persists beyond two weeks after birth, cholestasis or conjugated hyperbilirubinemia must be considered in the differential diagnoses. Neonatal cholestasis is defined as a direct bilirubin level of ≥ 2 mg/dl and ≥ 20% of the total bilirubin level. Persistent jaundice associated with acholic stools and dark urine may suggest biliary atresia. Such findings must undergo immediate diagnostic evaluation since the Kasai portoenterostomy surgical procedure for biliary atresia requires diagnosis at an early age.

Differential diagnoses include: Breastfed induced (insufficient production or intake of breast milk); physiologic (mild jaundice due to the immaturity of the liver); Gilbert Syndrome (a common benign genetic liver condition in which the liver doesn't properly process bilirubin and leads to isolated unconjugated hyperbilirubinemia); Crigler-Najjar syndrome (genetic disorder affecting metabolism of bilirubin); Rotor's syndrome (a benign, genetic liver disorder characterized by chronic, predominantly conjugated, non-hemolytic hyperbilirubinemia with normal liver histology); Dubin-Johnson syndrome (autosomal recessive disorder that causes an increase of conjugated bilirubin without elevation of liver enzymes); Biliary atresia (improper opening of bile ducts which leads to bile accumulation); Alpha-1 antitrypsin deficiency (genetic disorder that causes defective production of A1AT that protects the lungs and liver from damage); Hemolytic diseases (hereditary spherocytosis or G6PD deficiency); Hemoglobinopathies (Sickle cell anemia, Alpha/Beta Thalasemias); Congenial Hypothyroidism; Cystic fibrosis (results in abnormal bile production); Infections (Hepatitis, EBV, CMV, sepsis);

Hepatotoxins (APAP, antibiotics, anticonvulsants); or Vascular causes (Budd Chiari syndrome or Veno-occlusive disease).

Complications of hyperbilirubinemia causes concern since unconjugated bilirubin is neurotoxic and may lead to acute bilirubin encephalopathy, death, or lifelong neurologic sequelae. Serum unconjugated bilirubin levels greater than 30 mg/dl have been known to cause kernicterus. Clinical findings of kernicterus may consist of a sluggish Moro reflex, opisthotonos, hypotonia, vomiting, high-pitched cry, hyperpyrexia, seizures, paresis of gaze, oculogyric crisis, and death. Milder forms of bilirubin encephalopathy include cognitive dysfunction and learning disabilities.

Initial diagnostic evaluation for unconjugated hyperbilirubinemia may include: complete blood count, reticulocytes, direct and indirect Coombs test, haptoglobin, and Hb electrophoresis. Initial laboratory testing for conjugated hyperbilirubinemia consists of: CBC, liver enzymes, albumin, total protein, coagulation factors, cholesterol, and ammonia levels. Additional diagnostic tests include: TSH, Free T4, hepatitis and EBV serologies, serum alpha-1 antitrypsin levels, immunoglobulins, serum amino acids, and an EKG. Imaging studies directed toward diagnosis may begin with an abdominal ultrasound to measure hepatic size and/or consistency, and to detect abnormal echotexture as well as identifying masses, cysts, abscesses, and biliary tree abnormalities. New testing known as the Jaundice Chip Resequencing Array is available for inherited intrahepatic cholestasis of unknown origin. This test looks at several genes that are responsible for α-1 Antitrypsin deficiency, Alagille syndrome, FIC1 deficiency, BSEP deficiency, and MDR3 deficiency. Further imaging options include cholangiogram, CT, MRI, ERCP, and/or liver biopsies.

Treatment options for moderate to severe jaundice consist of: alteration of breast-feeding with formula feeding; Phototherapy to change the shape and structure of the bilirubin molecules in such a way that they can be excreted in the urine and stool; Intravenous immunoglobulin if caused by ABO or Rh isoimmunisation; Exchange transfusions for severe jaundice that doesn't respond to other treatments; Interruption of the enterohepatic circulation with medications such as Agar, Cholestyramine, activated charcoal, calcium phosphate; or Surgery if caused by biliary atresia or choledochal cysts.

Investigation of jaundice and timely treatment of the underlining pathology is crucial to patient outcomes. Age, medical history, and physical examination are key factors for accurate diagnosis. The providers at GI for Kids would be pleased to evaluate and help manage patients presenting with jaundice symptoms or assist with referral to the appropriate medical team.

Jaundice in Breast Fed Babies

Ashley Treadway, MS, RDN, LDN

Jaundice is more common in babies who are breastfed than babies who are formula fed. Breastfeeding jaundice usually occurs when a newborn does not get a good start on breastfeeding from either improper latch, inability to feed well, or when the child is supplemented with formula which can interfere with breastfeeding. Adequate amounts of breast milk will increase the baby's bowel movements, which will then help excrete the buildup of bilirubin.

If the bilirubin levels are less than 20mg/dl the following treatments are recommended:

• Initiate the breastfeeding relationship as soon as possible after birth.

• Increase feedings to 8-12 times a day.

• Feed whenever the baby is alert, sucking on the hands, and smacking the lips. This is one way babies let you know they are hungry. Do not try to put the baby on a "schedule," feed the baby frequently until the breastfeeding relationship is effectively established.

• Work with a lactation consultant to make sure the baby is latching on the breast well.

• It is perferable to breastfed, but if the serum bilirubin is eleveated and if the child is not intaking the appropriate volume then we will need to hold breastfeeding for 2-3 days and supplement with formula or supplement every other feed with formula to try to adequatley hydrate the child. If this decreases the serum bilirubin sufficiently we can try to restart breastfeeding.

• If supplementation is needed to increase the baby's intake, we encourage the mother to continue to pump during this time to not interrupt the production of her milk.

• Closely monitor baby's weight gain.

The baby should recover fully with the right monitoring and treatment. Although jaundice cannot be prevented, there are ways to avoid it becoming serious and reaching levels that require additional interventions, such as phototherapy and in severe cases, a blood transfusion.

References: www.americanpregnancy.com, www.kidshealth.org, www.nlm.nih.gov

JAUNDICE AT A GLANCE

Two types of jaundice may occur in newborns who are breastfed. Both types are most often harmless.

- Breastfeeding jaundice is seen in breastfed babies during the first week of life. It is more likely to occur when babies do not nurse well or the mother's milk is slow to come in.
- Breast milk jaundice may appear in some healthy, breastfed babies after day 7 of life. It is likely to peak during weeks 2 and 3 but may last at low levels for a month or more. The problem may be due to how substances in the breast milk affect the breakdown of bilirubin in the liver. Breast milk jaundice is different than breastfeeding jaundice.

Severe newborn jaundice may occur if the baby has a condition that increases the number of red blood cells that need to be replaced in the body, such as:

- Abnormal blood cell shapes
- Blood type mismatch between the mother and baby
- Bleeding underneath the scalp (cephalohematoma) caused by a difficult delivery
- Higher levels of red blood cells, which is more common in small-for-gestational-age babies and some twins
- Infection
- Lack (deficiency) of certain important proteins, called enzymes

Things that make it harder for the baby's body to remove bilirubin may also lead to more severe jaundice, including:

- Certain medicines
- Infections present at birth, such as rubella, syphilis, and others
- Diseases that affect the liver or biliary tract, such as cystic fibrosis or hepatitis
- Low oxygen level (hypoxia)
- Infections (sepsis)
- Many different genetic or inherited disorders

Our behavior health clinicians can help your pediatric patients with:

- **GI-related disorders** including recurrent abdominal pain, food allergies, celiac disease, irritable bowel syndrome & inflammatory bowel disease, constipation & encopresis
- **Other disorders** including adjustment to illness, family dysfunction, sleep difficulties, conduct disorder, depression, generalized anxiety, panic attacks, eating disorders, non-compliance to medical regimens, and more…

Visit our website for more details: www.GIforKids.com

Call for an appointment today!
865.546.3998

transitions
BEHAVIORAL HEALTH CLINIC

BEE FIT 4 KIDS

Bee Fit 4 Kids
www.BeeFit4Kids.com

- ☐ A one-on-one pediatric weight management program administered by a multi-disciplinary team.
- ☐ The format allows Registered Dietitians to identify nutrition and fitness trouble areas immediately. This helps the staff make appropriate changes so weight management success is maximized.

Visit our website, www.BeeFit4Kids.com, for more details, or call 865.546.3998.

Now accepting insurance!

East Tennessee Children's Hospital
Gastroenterology and Nutritional Services
Medical Office Building
2100 Clinch Avenue, #510
Knoxville, TN 37916

PRSRT STD
U.S. POSTAGE
PERMIT NO. 433
KNOXVILLE, TN

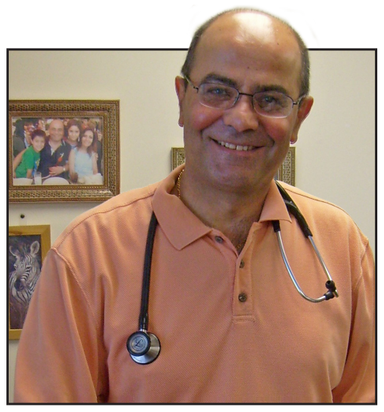
GI for Kids, PLLC

East Tennessee Children's Hospital
Gastroenterology and Nutrition Services

Fall/Winter 2015 www.giforkids.com Volume 6 Issue 8

Friends and Colleagues,

As the seasons are changing and we are approaching winter, we at GI for Kids hope for a great holiday season for you and your family. Our focus for this issue is jaundice in infants and children. Hyperbilirubinemia resulting in jaundice is one of the most common problems encountered in term newborns. Few will have serious underlying pathology. However, hyperbilirubinemia in the newborn period can be associated with severe illnesses such as hemolytic disease, anatomic abnormalities of the liver, metabolic and endocrine disorders and infections. The management goals at GI for Kids are to exclude pathologic causes of hyperbilirubinemia and initiate treatment to prevent bilirubin neurotoxicity. Please contact us with any questions or referrals and visit our website at www.giforkids.com for more information on all of the pediatric services our practice provides.



Youhanna S. Al-Tawil, MD
Medical Director

Celi-Act

Celiac Disease, or gluten intolerance, is a genetic autoimmune disease estimated to affect 1 of every 133 people in the U.S. Research indicates approximately 2.5 million people in the U. S. have celiac disease with approximately 80,000 diagnosed.

East Tennessee Children's Hospital

CELI-ACT IS SPONSORED BY GI FOR KIDS, PLLC, LOCATED AT EAST TENNESSEE CHILDREN'S HOSPITAL, AND OPEN TO ANYONE WITH CELIAC DISEASE OR THEIR FAMILY MEMBERS.

Celi-Act
Knoxville area celiac patients helping each other.
www.celi-act.com