

MINISTRY OF HEALTH OF UKRAINE
Ukrainian Medical Stomatological Academy

«Approved»
on meeting the
department of Pediatric Surgery
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METHODICAL INSTRUCTIONS

FOR STUDENTS' SELF-WORK

WHILE PREPARING FOR PRACTICAL LESSONS

<i>Educational discipline</i>	Pediatric Surgery
<i>module №5</i>	Congenital Anomalies in Children
<i>Theme of the lesson</i>	Malformations of the liver and bile ducts, biliary atresia
<i>Course</i>	VI
<i>Faculty</i>	foreign students preparation

POLTAVA 2020

1.The topic basis: Actuality of theme of employment is conditioned plenty of diagnostic errors, complications and unsatisfactory consequences of treatment.

The topic “ Malformations of the liver and bile ducts, biliary atresia” is very important for future doctors in their professional activity, positively influences the students in their attitude to the future profession, forms professional skills and experience as well as taking as a principle the knowledge of the subject learnt

2.The aims of the training course:

1. A student must have general presentations about the basic stages of embryogenesis of the liver and bile ducts
2. A student must know:
 - Ø Classifications of malformations of liver and bile ducts
 - Ø Clinical displays, features of clinical picture depending on a level and type of malformations
 - Ø Modern methods of diagnostics at the malformations of liver and bile ducts, principles of differential diagnostics
 - Ø Basic questions of treatment, principles of grant a help on the different stages of treatment
3. A student must seize:
 - Ø By the method of review of children with the malformations of liver and bile ducts
4. A student must be able:
 - Ø to diagnose the malformations of liver and bile ducts
 - Ø to distinguish the features of clinical picture depending on a level and type of malformations of liver and bile ducts
 - Ø to conduct differential diagnostics
 - Ø to analyse and estimate the results of laboratory and instrumental researches
 - Ø to appoint therapy a child with the malformations of liver and bile ducts on the different stages of treatment
 - Ø to estimate efficiency of the conducted therapy.
5. A student must develop the creative capabilities:
 - Ø in the process of clinical inspection and treatment of children with the malformations of liver and bile ducts, conducting research in obedience to by the conducted department by research work
 - Ø to conduct the scientific analysis of sources on this issue

3.Basic knowledge, skills, habits necessary for studying the subject (interdisciplinary integration).

Names of previous disciplines	Obtained skills
1. Anatomy, operative Surgery, Topografic Anatomy	Anatomy of the abdominal cavity organs, topographic anatomy of the abdominal cavity, embryogenesis of the digestive system. To establish a possible developmental defect, to determine the places of operative access
2. Physiology.	Anatomical and physiological properties of the digestive system

3. Pathological Physiology	Indicators of a general analysis of blood, urine, biochemical blood test, coagulogram of acid-base state in norm and pathology. To distinguish between normal and pathological indicators, to interpret changes.
4. Surgical diseases, operative Surgery and topographic anatomy.	Principles of care for surgical patients. Clinic, diagnosis, treatment of intestinal obstruction. Measure CVP, establish gastric tube, urinary catheter, catheterize veins
5. Propedeutics of childhood diseases Faculty and hospital pediatrics, neonatology	Symptoms of liver and bile duct pathology Measure blood pressure, heart rate, the number of respiratory movements per minute, conduct a survey of the abdomen (palpation, percussion, auscultation), rectal examination
6. Pharmacology and Clinical Pharmacology	Pharmacology, pharmacodynamics and pharmacokinetics of drug groups that are used in the surgical pathology of abdominal organs in infants Calculate doses of drugs in the provision of emergency care and different routes of administration
7. Radiology	Sonographic anatomy of the abdominal cavity is normal. Determine the size, structure, abnormalities of the liver and biliary tract with ultrasound

Theoretical questions for the lesson:

1. Determination of reasons of malformations of liver and bile ducts
2. Basic stages of embryogenesis of liver and bile ducts
3. Classifications of imperforations of bile ducts
4. Clinic of malformations of liver and bile ducts for babies
5. Diagnostics of malformations of liver and bile ducts
6. Treatment of malformations of liver and bile ducts

4. Maintenance of the subject:

Biliary Atresia - absence or malformation of the [BILE DUCTS](#), also called neonatal CHOLESTASIS. Biliary atresia is nearly always congenital (present at birth). In some infants biliary atresia appears to develop as a consequence of an inflammatory process that occurs shortly after birth, destroying the bile ducts. Biliary atresia prevents the flow of [BILE](#) from the [LIVER](#), causing toxins to accumulate in the liver.

Symptoms of Biliary Atresia

Symptoms depend on the extent of the atresia and may become apparent within days of birth or manifest slowly over the first six months of life. Symptoms include

- [JAUNDICE](#), a yellowish discoloration of the [SKIN](#) resulting from the liver's inability to break down BILIRUBIN into components the body can excrete as waste
- stools that are pale in color, the consistency of clay, and unusually foul smelling
- dark [URINE](#)
- failure to grow or gain weight
- [ABDOMINAL DISTENTION](#) resulting from enlarged [SPLEEN](#)
- fussiness and irritability

Neonatal Jaundice

NEONATAL JAUNDICE is fairly common, affecting about 50 percent of full-term and 80 percent of preterm (premature) newborns. It points to biliary atresia or other LIVER conditions only when it is apparent within the first 24 hours after birth or persists despite treatment.

The diagnostic path includes blood tests to measure the amounts of bilirubin in the blood and [PERCUTANEOUS LIVER BIOPSY](#) to determine whether the hepatocytes, the cells that process bilirubin, are normal. Normal hepatocytes strongly suggest biliary atresia. Other diagnostic procedures may include ULTRASOUND and intraoperative cholangiography (injecting dye into the bile ducts to visualize them using FLUOROSCOPY or other imaging technologies).

Biliary Atresia Treatment

The only treatments for biliary atresia are surgical procedures to help restore the flow of bile. The first of these procedures is hepatic portoenterostomy, in which the surgeon creates an opening between the [JEJUNUM](#) (middle segment of the [SMALL INTESTINE](#)) and the bile duct structures that exist outside the liver. This procedure allows bile to drain directly from the liver into the small intestine and can achieve adequate liver function for up to several years. However, it does not correct the structural defects of the bile transport network within the liver, and damage to the liver (fibrosis and CIRRHOSIS) continues. Nearly all infants who have biliary atresia require [LIVER TRANSPLANTATION](#), the second surgical procedure to treat the condition, before they are three years old. Long-term success of liver transplantation depends on numerous variables.

Extrahepatic biliary atresia is a rare gastrointestinal disorder characterized by destruction or absence of all or a portion of the bile duct that lies outside the liver (extrahepatic bile duct). The bile duct is a tube that allows the passage of bile from the liver into the gall bladder and, eventually, the small intestine. Bile is a liquid secreted by the liver that plays an essential role in carrying waste products from the liver, breaking down fats in the small intestine, and promoting absorption of vitamins. In extrahepatic biliary atresia, absence or destruction of the bile ducts results in the abnormal accumulation of bile in the liver. Affected infants have yellowing of the skin and whites of the eyes (jaundice) and scarring of the liver (cirrhosis). In some cases, additional abnormalities may be present, including heart defects and kidney and spleen malformations. The exact cause of extrahepatic biliary atresia is unknown.

Symptoms

The symptoms of extrahepatic biliary atresia are usually obvious by the age of two to four weeks and include a yellowish coloration of the skin and whites of the eyes (jaundice), abnormally pale stools, dark urine, swollen (distended) stomach, and/or abnormal enlargement of the liver (hepatomegaly). By the age of six to 10 weeks, additional symptoms may also develop including itchiness (pruritis), irritability, growth delays, and/or an increase in blood pressure within the veins that carry blood from the intestine to the liver (portal hypertension). Bile ducts inside the liver (intrahepatic bile ducts) are also involved. If left untreated, biliary atresia may result in scarring of the liver (cirrhosis) and, eventually, liver (hepatic) failure. In some cases, children with biliary atresia may have additional congenital abnormalities including malformations of the heart (e.g., situs inversus, levocardia, and ventricular septal defects) and/or kidneys. Situs inversus is a condition in which the internal organs are on the opposite side of the body from normal. Levocardia is a condition in which the heart is malpositioned. (For more information on ventricular septal defects see the Related Disorders section below.) Additional features may be associated with some cases of extrahepatic biliary atresia including absence of the spleen (asplenia), the presence of more than one spleen (polysplenia), and/or other anatomical abnormalities.

Causes

The exact cause of extrahepatic biliary atresia is unknown. Several factors may contribute to the development of the disorder, including immunologic, infectious, genetic, environmental, and/or other factors. In most cases of extrahepatic biliary atresia, the bile ducts are normal at birth, but something causes them to be damaged and replaced with fibrous tissue (sclerosis). Several viruses,

including cytomegalovirus, reovirus type 3 and rotavirus infections are being studied as possible causative agents.

A minority of cases may be caused by defects during the development (morphogenesis) of the liver and biliary tree during pregnancy. Biliary atresia is not an inheritable disease, and no specific genetic predisposition for the disorder has been reported.

Affected Populations

Extrahepatic biliary atresia is a rare disorder with a slight increased frequency in females. It occurs in approximately 1 in 10,000 to 15,000 births in the United States. Approximately 400-600 new cases of biliary atresia are encountered in the United States each year. According to one estimate, the prevalence of biliary atresia in Europe is approximately 1 in 12,000 births. Biliary atresia is the most common cause of chronic liver disease in children.

Related Disorders

Symptoms of the following disorders can be similar to those of extrahepatic biliary atresia. Comparisons may be useful for a differential diagnosis: Neonatal hepatitis (sometimes referred to as neonatal cholestasis) is a term used to describe a group of disorders that present with jaundice due to inflammation of the liver and/or injury of the bile duct structure within the liver (intrahepatic). Some of the more common disorders are the Alagille syndrome, deficiency of alpha-1-antitrypsin protein, cystic fibrosis, progressive familial intrahepatic cholestasis, and defects in synthesis of bile acids. The cause for neonatal hepatitis in some infants remains unknown. Some studies suggest an association with an infectious or viral disease. Symptoms of neonatal hepatitis typically appear during the first few weeks of life and may include a yellow discoloration of the skin and whites of the eyes (jaundice), an abnormally enlarged liver (hepatomegaly), poor feeding, slow growth, red itchy skin, abdominal discomfort, lightly colored stools, and/or dark urine. (For more information on this disorder, choose "Neonatal Hepatitis" as your search term in the Rare Disease Database.)

Primary sclerosing cholangitis is a rare progressive disorder characterized by inflammation, thickening, and abnormal formation of fibrous tissue (fibrosis) within the passages that carry bile from the liver (bile ducts). This often results in the decrease of bile flow from the liver (cholestasis). This disorder is more common in older children and adults. The neonatal form of primary sclerosing cholangitis is rare and may share features with biliary atresia and neonatal hepatitis. The cause is not known, but most cases are associated with defects in the immunologic or hematologic systems. Symptoms associated with primary sclerosing cholangitis include fatigue and itching (pruritis), followed by yellowing of the skin, mucous membranes, and whites of the eyes (jaundice). In addition, affected individuals may have dark urine, light-colored stools, abdominal pain, and/or nausea. The exact cause of primary sclerosing cholangitis is not known. (For more information on this disorder, choose "primary sclerosing cholangitis" as your search term in the Rare Disease Database.)

The following disorders may be associated with extrahepatic biliary atresia as secondary characteristics. They are not necessary for a differential diagnosis: Ventricular septal defects (VSDs) are heart defects that are present at birth (congenital). The normal heart has four chambers. The two upper chambers, known as atria, are separated from each other by a fibrous partition known as the atrial septum. The two lower chambers are known as ventricles and are separated from each other by the ventricular septum. Valves connect the atria (left and right) to their respective ventricles. The aorta, the main vessel of arterial circulation, carries blood from the left ventricle and away from the heart. VSDs can occur in any portion of the ventricular septum. The size and location of the defect determine the severity of the symptoms. Small ventricular septal defects can close on their own (spontaneously) or become less significant as the child matures and grows. Moderately-sized defects can cause congestive heart failure, which is characterized by an abnormally rapid rate of breathing (tachypnea), wheezing, unusually fast heartbeat (tachycardia), enlarged liver (hepatomegaly), and/or failure to thrive. (For more information on this disorder, choose "ventricular septal defects" as your search term in the Rare Disease Database.)

Standard Therapies

Diagnosis

The diagnosis of extrahepatic biliary atresia requires a direct examination of the bile ducts by abdominal surgery (laparotomy) and the microscopic examination of tissue from the liver (liver biopsy). During the surgery special contrast dye is injected into the common bile duct and x-ray films are taken to outline the major bile ducts (direct cholangiogram). These films show the movement (or lack of movement) of the dye through the liver, bile ducts, and small intestine. The physician is then able to evaluate the structure of the bile ducts and to determine the site of the blockage (proximal or distal). Blood tests may demonstrate elevated levels of liver enzymes and bilirubin and detect viral or bacterial agents. Ultrasound of the liver may show absence of the gall bladder.

Treatment

No cure exists for biliary atresia, but the timely diagnosis and surgical intervention improves short- and long-term outcomes in most patients. Special attention to the nutritional needs and diet are essential for children with this disorder. Special supplements, formulas, and dietary restrictions may be necessary for affected infants. Surgery must be performed to remove the obstruction and allow bile to flow into the bile ducts and small intestine (Kasai hepatoportoenterostomy). In this procedure, the extrahepatic bile ducts are removed and replaced with a portion of the affected infant's small intestine thereby restoring normal bile flow. The exact surgical procedure may vary according to the location and nature of the obstruction. In the majority of cases, bile flow can be established with these surgical procedures. However, some children may experience variable degrees of liver dysfunction even after successful surgery. The Kasai procedure may also be used as an early intermediate procedure to support the child's growth. Despite the Kasai procedure, liver transplantation may ultimately become necessary in many cases. Antibiotics may be used to treat infections of the bile ducts (cholangitis).

Genetic counseling may be of benefit for people with extrahepatic biliary atresia and their families. Other treatment is symptomatic and supportive.

Liver Cysts

Cysts can be divided into benign (non-cancerous) congenital cysts (present at birth) or acquired cysts; that is, a new cyst caused from a traumatic injury, for instance. Cysts present at birth may result from abnormally developed bile ducts. Physicians use the term solitary cyst to refer to liver cysts that are not associated with the inherited polycystic liver disease, even though solitary cysts may be multiple in nature. Polycystic (having many cysts) liver disease can affect the liver, kidneys, lung and pancreas. Additionally, some patients are prone to neoplastic cysts, where the liver tissue grows abnormally, forming a mass that can either be benign (non-cancerous) or malignant (cancerous).

Sometimes surgeons can treat congenital or acquired cysts by aspirating, or using suction, to remove the fluid. In other cases it is necessary to surgically remove them and this can usually be done laparoscopically.

Benign Liver Masses

Many disease areas in the liver may appear as single or multiple liver masses that are benign (non-cancerous). The most typical benign liver masses include cavernous hemangioma, hepatocellular adenomas and focal nodular hyperplasia.

Cavernous hemangioma is one of the more common liver tumors. Tangled and multiple blood vessels, supported by walls of collagen (the fibrous protein found in cartilage, bone and tendon), characterize this benign tumor, generally found more in women than men. It is very rare that a hemangioma will burst and they never become cancerous. If the hemangioma is large and causing discomfort, then it is surgically removed.

Hepatocellular adenomas are non-cancerous liver tumors typically found in women who use birth control pills. Adenomas can also occur in men who have a history of steroid use. Because of the risk of bleeding, rupture and the small chance that the tissue could turn cancerous, surgery is

the treatment of choice for most large adenomas in patients who are suitable surgical candidates. Small adenomas may be observed and monitored in some cases.

Focal nodular hyperplasia (FNH) is a highly common, benign tumor of the liver. The risk for bleeding or the development of cancer has not been documented. Treatment depends on the certainty of the diagnosis, as the most difficult challenge is to distinguish FNH from cancerous liver tumors or adenomas. If it is not possible to definitively rule out a cancer, then surgery is the safer choice.

Primary Malignancies of the Liver, Gallbladder and Biliary Tree

Primary (meaning they start in the liver) malignant (cancerous) tumors of the liver include hepatocellular carcinoma, cholangiocarcinoma, hepatoblastoma, and a number of other less frequently occurring cancerous tumors.

Hepatocellular carcinoma (HCC) accounts for the overwhelming majority of primary liver cancer and Hepatitis B (HBV) is the leading cause of HCC worldwide. Hepatitis C (HCV) is also a common cause of HCC in the United States, but all patients with cirrhosis of the liver are at increased risk for developing HCC. Surgical resection to remove the tumor is the best treatment depending on the extent of disease. Survival rates are better when tumors are small and can be completely removed. Patients who have severe liver disease and portal hypertension may not be candidates for surgical resection, but may be best treated by liver transplantation. However, when tumors invade both lobes of the liver (multifocal), have spread outside the liver (for example, to the lungs), or in cases where the tumor in the liver originated from some other site in the body and then spread to the liver, surgery is generally not recommended. In this case, other treatment options include:

- Chemotherapy - either systemic (treating the entire system) or intra-arterial (delivering the chemotherapy through a catheter directly into the artery supplying the liver)
- Radiofrequency ablation (placing a probe into the tumor and heating up the tumor using radio waves in order to shrink/destroy the cancerous tissue)
- Chemoembolization (injecting small particles containing a chemotherapy drug into the hepatic artery to block blood supply to the tumor and shrink/destroy the cancerous tissue)
- Radiation therapy (targeting the tumor with X-rays or radioactive agents)
- Cyberknife® (precisely targeted radiation therapy, beamed at the tumor site, with minimal exposure to surrounding healthy tissue); Cyberknife® is especially well suited for hard-to-reach and complex tumors in the liver and pancreas, among other organs.

Physicians continue to evaluate newer non-surgical approaches to treat HCC.

Normally the liver can regenerate (grow back) after a portion has been removed, but this ability is limited if you have cirrhosis (or scarring) of the liver. As mentioned above, if you also have extensive HCC that cannot be removed surgically, or that does not respond to other treatment procedures, you may be a candidate for liver transplantation.

Cholangiocarcinoma is cancer of the bile duct system (the biliary tree). This can arise in the liver from the bile duct cells or in the major duct draining bile from the liver called the common bile duct. A slow growing, rare type of cancer that often occurs at the junction of the right and left hepatic ducts as they leave the liver (Klatskin's tumor), surgical resection is the treatment of choice. If the tumor cannot be removed surgically, liver transplantation may be an option for some selected patients, and for other patients treatment includes chemotherapy and radiation.

Gallbladder carcinoma is an uncommon cancer that may arise anywhere in the gallbladder and frequently invades into the adjacent liver tissues. Surgical treatment offers the best hope for cure if the tumor has not spread elsewhere and can be resected. This includes removing the gallbladder, the surrounding liver tissue, and the lymph nodes around the liver. Sometimes the common bile duct must be removed to achieve clear surgical margins.

Hepatoblastoma is a highly malignant tumor that occurs almost exclusively in children less than five years of age, although older children and adults have been diagnosed with this condition as well. A large, single tumor that is usually found in the right lobe of the liver, hepatoblastomas are generally associated with abnormalities at birth. Surgery to remove the tumor is the preferred

treatment. Radiation and/or chemotherapy before surgery may benefit some patients. If the tumor is not surgically removable, liver transplantation may be an option.

Metastatic Malignancies of the Liver

Metastases are cancer cells that have spread from an original or primary site to one or more locations or organs elsewhere in the body. For example, cancer that begins in the colon (primary) can metastasize to the liver. The liver is a prime site for metastatic cancer for many reasons; chiefly, the liver receives 80 percent of its blood supply from the portal vein that receives the blood draining from the many primary cancer sites, such as the colon and rectum.

The most common primary tumors that cause liver metastases include lung, colon, pancreas, breast, stomach, ovary, prostate, gallbladder and cervix. If there is not evidence of other systemic disease or if the systemic disease is well-controlled, surgical resection is the treatment of choice if all the tumor can be removed and still leave an adequate amount of liver tissue. If surgery is not an option then chemotherapy, radiofrequency ablation, chemoembolization, or Cyberknife® may be used.

Portal Hypertension

Portal hypertension is a common complication of cirrhosis, which in turn may be caused by many different liver diseases in which normal, healthy tissue is replaced with nonfunctioning "scar" tissue, and healthy, functioning liver cells are lost. With portal hypertension, scar tissue prevents the vein that drains blood from the spleen and intestines (portal vein) from bringing the blood through the liver. This condition creates increased pressure in the veins, which can lead to other problems, such as fluid buildup (ascites) in the abdomen and elsewhere, and enlarged veins (varices) in the esophagus or stomach that can bleed. Treatment options to manage complications from portal hypertension include the following.

- **Endoscopic treatment in the Center for Interventional Gastroenterology:** Gastroenterologists (physicians who treat disorders of the digestive tract) use a lighted, flexible tube (introduced through the mouth) to visualize, diagnose and stop bleeding varices by using rubber bands to block the vessels ("banding") or by injecting a solution into the varices so they scar ("sclerotherapy").

- **Medication:** Physicians may prescribe medication (separately or together with endoscopic therapy) to lower variceal pressure and reduce the likelihood of bleeding.

- **Diet:** It is important to follow a low sodium diet (typically 2 grams/day) to avoid fluid collection (ascites) and minimize edema (swelling). Our nutritionist can explain which foods are high in sodium, what alternatives and substitutes you might consider, and how to interpret food labels. Also, to help prevent encephalopathy (confusion, drowsiness and behavioral changes brought on by liver disease), you must take your medications exactly as directed. Protein is not restricted in the case of encephalopathy; however, the type of protein you eat is important. For example, our nutritionist may advise you to eat a mix of protein foods from meat and vegetable sources such as eggs, dairy products, nuts, lentils and beans, but to consume less red meat. Also, you may be able to better tolerate protein-rich foods if you eat them throughout the day rather than all at once. Click [here](#) to read more about diet and nutrition.

- **Specialized Portal Decompressive Surgery:** If physicians cannot control bleeding varices with endoscopic therapy or medication, then surgeons at the Transplant Institute perform highly specialized portal decompressive surgery to reconfigure blood flow through the liver. Portal decompressive surgery, which can include portacaval or other shunting procedures, reduces pressure in the veins draining the liver, and in so doing, decreases the swelling in the variceal veins to stop the bleeding and lessen the risk of rupture. These procedures are done by sewing a vein such as the portal vein or splenic vein, which are under high pressure, to a vein under low pressure.

TIPS, or Transjugular Intrahepatic Portosystemic Shunt, is a specialized procedure used to treat portal hypertension. Radiologists insert a metal stent in the liver, via the jugular vein in the neck. The stent (a tiny wire mesh tube), creates a passageway between the portal and hepatic veins to promote blood flow through the liver from the portal vein directly to the hepatic vein thereby

bypassing the blockage caused by the scarring of cirrhosis and relieving the high pressure in the liver. However, some patients may have vein abnormalities (such as a clot in the portal vein) that prohibit the TIPS procedure or may be at greater risk for mental confusion (encephalopathy) following the procedure.

The Budd-Chiari Syndrome, an uncommon syndrome characterized by obstruction of veins draining blood out of the liver, can also cause portal hypertension. Some patients are born with a membrane-type web that actually blocks blood flow. Patients experience abdominal pain, ascites (fluid collection in the abdomen) and abnormal liver function. A radiology exam confirms the diagnosis by highlighting blockage in the hepatic veins. Most patients need surgery to improve, either to repair the membranous webs or to perform a side-to-side portacaval shunt procedure (sewing the portal vein to the inferior vena cava) to reduce the high blood pressure in the liver.

What are congenital liver defects?

Defects of the liver at birth usually affect the bile ducts. Though rare, some congenital liver defects include the following:

- **Biliary atresia.** A condition in which the bile ducts are blocked or have developed abnormally to obstruct flow of bile in infants.
- **Choledochal cyst.** A malformation of the hepatic duct that can obstruct flow of bile in infants.

What are the indicators that a congenital liver defect may be present?

Congenital liver defects that affect the flow of bile share some common symptoms. The following are the most common symptoms of congenital liver defect. However, each individual may experience symptoms differently. Symptoms may include:

- Jaundice - yellowing of the skin and eyes
- Dark urine
- Pale, white or gray-colored stools

The symptoms of congenital liver defects may resemble other medical conditions or problems. Always consult your child's physician for a diagnosis.

How are congenital liver defects diagnosed?

Congenital liver defects that affect the flow of bile are usually diagnosed at birth or shortly afterward. In addition to a complete medical history and physical examination, diagnostic procedures for a congenital liver defect may include the following:

- **Laboratory tests (blood, urine and stool).**
- **Liver function tests.** A series of special blood tests that can determine if the liver is functioning properly.
- **Liver biopsy.** A procedure in which tissue samples from the liver are removed (with a needle or during surgery) from the body for examination under a microscope.
- **Computed tomography scan (CT or CAT scan).** A diagnostic imaging procedure using a combination of X-rays and computer technology to produce cross-sectional images (often called slices), both horizontally and vertically, of the body. A CT scan shows detailed images of any part of the body, including the bones, muscles, fat, and organs. CT scans are more detailed than general X-rays.
- **Ultrasound (also called sonography).** A diagnostic imaging technique, which uses high-frequency sound waves and a computer to create images of blood vessels, tissues, and organs. Ultrasounds are used to view internal organs of the abdomen such as the liver, spleen, and kidneys and to assess blood flow through various vessels.

What is the treatment for congenital liver defects?

Specific treatment for congenital liver defects will be determined by your child's physician based on:

- Your child's age, overall health, and medical history
- Extent of the disease
- Your child's tolerance for specific medications, procedures, or therapies
- Expectations for the course of the disease

- Your opinion or preference

Treatment may include surgery to reconstruct or bypass the bile ducts. Sometimes, a liver transplant may be necessary.

5. Additional materials for the self-control

A. Clinical cases

Case 1.

A one-month child has an icterus which appeared since the first day and gradually becomes more intensive, acquiring a green tint. The excrement is colourless since the child's birth, the urine is of a dark beer color. The enlargement of the liver is detected. The blood analysis showed hyperbilirubinemia due to direct bilirubin. In the urine having bilious pigments the urobilin is absent. Your diagnosis?

B. Tests

6. LITERATURE FOR STUDENTS

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