

## MEDICAL POLICY

<b>POLICY TITLE</b>	<b>LIVER TRANSPLANT AND COMBINED LIVER-KIDNEY TRANSPLANT</b>
<b>POLICY NUMBER</b>	<b>MP 9.006</b>

<b>CLINICAL BENEFIT</b>	<input type="checkbox"/> MINIMIZE SAFETY RISK OR CONCERN. <input type="checkbox"/> MINIMIZE HARMFUL OR INEFFECTIVE INTERVENTIONS. <input type="checkbox"/> ASSURE APPROPRIATE LEVEL OF CARE. <input type="checkbox"/> ASSURE APPROPRIATE DURATION OF SERVICE FOR INTERVENTIONS. <input checked="" type="checkbox"/> ASSURE THAT RECOMMENDED MEDICAL PREREQUISITES HAVE BEEN MET. <input type="checkbox"/> ASSURE APPROPRIATE SITE OF TREATMENT OR SERVICE.
<b>Effective Date:</b>	<b>3/1/2024</b>

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### I. POLICY

A liver transplant, using a cadaver or living donor, is **medically necessary** for carefully selected individuals with end-stage liver failure due to irreversibly damaged livers.

Etiologies of end-stage liver disease include, but are not limited to, the following:

- Hepatocellular diseases
  - Alcoholic cirrhosis
  - Viral hepatitis (either A, B, C, or non-A, non-B)
  - Autoimmune hepatitis
  - Alpha-1 antitrypsin deficiency
  - Hemochromatosis
  - Non-alcoholic steatohepatitis
  - Protoporphyrria
  - Wilson disease
- Cholestatic liver diseases
  - Primary biliary cirrhosis
  - Primary sclerosing cholangitis with development of secondary biliary cirrhosis
  - Biliary atresia
- Vascular disease
  - Budd-Chiari syndrome
- Primary hepatocellular carcinoma (see Policy Guidelines section for patient selection criteria)
- Inborn errors of metabolism
- Trauma and toxic reactions
- Miscellaneous
  - Familial amyloid polyneuropathy

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Liver transplantation may be considered **medically necessary** in individuals with polycystic disease of the liver who have massive hepatomegaly causing obstruction or functional impairment.

Liver transplantation may be considered **medically necessary** in individuals with unresectable hilar cholangiocarcinoma (see Policy Guidelines for member selection criteria).

Liver transplantation may be considered **medically necessary** in pediatric individuals with nonmetastatic hepatoblastoma.

Liver *retransplantation* may be considered **medically necessary** in individuals with:

- primary graft nonfunction
- hepatic artery thrombosis
- chronic rejection
- ischemic type biliary lesions after donation after cardiac death
- recurrent non-neoplastic disease causing late graft failure

Combined liver-kidney transplantation may be considered **medically necessary** in individuals who qualify for liver transplantation and have advanced irreversible kidney disease.

Liver transplantation is considered **investigational** in the following individuals as there is insufficient evidence to support a general conclusion concerning the health outcomes or benefits associated with this procedure:

- Individuals with an intrahepatic cholangiocarcinoma
- Individuals with neuroendocrine tumors metastatic to the liver

Liver transplantation is considered **not medically necessary** in the following members:

- Individuals with hepatocellular carcinoma that has extended beyond the liver (see Policy Guidelines section for patient selection criteria)
- Individuals with ongoing alcohol and/or drug abuse. (Evidence for abstinence may vary among liver transplant programs, but generally a minimum of 3 months is required.)

Liver transplantation is considered **investigational** in all other situations not described above as there is insufficient evidence to support a general conclusion concerning the health outcomes or benefits associated with this procedure.

## POLICY GUIDELINES

### Contraindications

Potential contraindications subject to the judgment of the transplant center:

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1. Known current malignancy, including metastatic cancer
2. Recent malignancy with high risk of recurrence
3. Untreated systemic infection making immunosuppression unsafe, including chronic infection
4. Other irreversible end-stage disease not attributed to liver disease
5. History of cancer with a moderate risk of recurrence
6. Systemic disease that could be exacerbated by immunosuppression
7. Psychosocial conditions or chemical dependency affecting ability to adhere to therapy

### LIVER SPECIFIC PATIENT SELECTION CRITERIA

The Model for End-stage Liver Disease (MELD) and Pediatric End-stage Liver Disease (PELD) scores range from 6 (less ill) to 40 (gravely ill). The MELD and PELD scores will change during the course of a patient's tenure on the waiting list.

Patients with liver disease related to alcohol or drug abuse must be actively involved in a substance abuse treatment program.

Tobacco consumption is a contraindication.

Patients with polycystic disease of the liver do not develop liver failure but may require transplantation due to the anatomic complications of a hugely enlarged liver. The MELD and PELD score may not apply to these cases. One of the following complications should be present:

- Enlargement of liver impinging on respiratory function
- Extremely painful enlargement of liver
- Enlargement of liver significantly compressing and interfering with function of other abdominal organs

Patients with familial amyloid polyneuropathy do not experience liver disease, per se, but develop polyneuropathy and cardiac amyloidosis due to the production of a variant transthyretin molecule by the liver. MELD and PELD exception criteria and scores may apply to these cases. Candidacy for liver transplant is an individual consideration based on the morbidity of the polyneuropathy. Many patients may not be candidates for liver transplant alone due to coexisting cardiac disease.

### Hepatocellular Carcinoma

Criteria used for patient selection of hepatocellular carcinoma patients eligible for liver transplant include the Milan criteria, which is considered the criterion standard, the University of California, San Francisco (UCSF) expanded criteria, and United Network of Organ Sharing (UNOS) criteria.

**Milan criteria:** a single tumor 5 cm or less in diameter or 2 to 3 tumors 3 cm or less.

**University of California, San Francisco expanded criteria:** a single tumor 6.5 cm or less or up to 3 tumors 4.5 cm or less, and a total tumor size of 8 cm or less.

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**UNOS T2 criteria:** A single tumor 2 cm or greater and up to 5 cm or less or 2 to 3 tumors 1 cm or greater and up to 3 cm or less and without extrahepatic spread or macrovascular invasion. United Network for Organ Sharing criteria were updated in 2022.

Patients with HCC are appropriate candidates for liver transplant only if the disease remains confined to the liver. Therefore, the patient should be periodically monitored while on the waiting list, and if metastatic disease develops, the patient should be removed from the transplant waiting list. In addition, at the time of transplant, a backup candidate should be scheduled. If locally extensive or metastatic cancer is discovered at the time of exploration prior to hepatectomy, the transplant should be aborted, and the backup candidate scheduled for transplant.

Note that liver transplantation for those with T3 HCC is not prohibited by UNOS guidelines, but these patients do not receive any priority on the waiting list. All patients with HCC awaiting transplantation are reassessed at 3-month intervals. Those whose tumors have progressed and are no longer T2 tumors will lose the additional allocation points.

Additionally, nodules identified through imaging of cirrhotic livers are given a Class 5 designation. Class 5B and 5T nodules are eligible for automatic priority. Class 5B criteria consist of a single nodule 2 cm or larger and up to 5 cm (T2 stage) that meets specified imaging criteria. Class 5T nodules have undergone subsequent locoregional treatment after being automatically approved on initial application or extension. A single Class 5A nodule (greater than 1 cm and less than 2 cm) corresponds to T1 HCC and does not qualify for automatic priority. However, combinations of Class 5A nodules are eligible for automatic priority if they meet stage T2 criteria. Class 5X lesions are outside of stage T2 and are not eligible for automatic exception points. Nodules less than 1 cm are considered indeterminate and are not considered for additional priority. Therefore, the UNOS allocation system provides strong incentives to use locoregional therapies to downsize tumors to T2 status and to prevent progression while on the waiting list.

**Cholangiocarcinoma**

According to the Organ Procurement and Transplantation Network (OPTN) policy on liver allocation, candidates with cholangiocarcinoma (CCA) meeting the following criteria will be eligible for a MELD or PELD exception with a 10% mortality equivalent increase every 3 months:

- Centers must submit a written protocol for patient care to the OPTN and UNOS Liver and Intestinal Organ Transplantation Committee before requesting a MELD score exception for a candidate with cholangiocarcinoma. This protocol should include selection criteria, administration of neoadjuvant therapy before transplantation, and operative staging to exclude patients with regional hepatic lymph node metastases, intrahepatic metastases, and/or extrahepatic disease. The protocol should include data collection as deemed necessary by the OPTN/UNOS Liver and Intestinal Organ Transplantation Committee.
- Candidates must satisfy diagnostic criteria for hilar CCA: malignant-appearing stricture on cholangiography and one of the following: carbohydrate antigen 19-9 100 U/mL, or and biopsy or cytology results demonstrating malignancy, or aneuploidy. The tumor

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should be considered unresectable on the basis of technical considerations or underlying liver disease (e.g., primary sclerosing cholangitis).

- If cross-sectional imaging studies (computed tomography [CT] scan, ultrasound, magnetic resonance imaging [MRI]) demonstrate a mass, the mass should be 3 cm or less.
- Intra- and extrahepatic metastases should be excluded by cross-sectional imaging studies of the chest and abdomen at the time of initial exception and every 3 months before score increases.
- Regional hepatic lymph node involvement and peritoneal metastases should be assessed by operative staging after completion of neoadjuvant therapy and before liver transplantation. Endoscopic ultrasound-guided aspiration of regional hepatic lymph nodes may be advisable to exclude patients with obvious metastases before neoadjuvant therapy is initiated.
- Transperitoneal aspiration or biopsy of the primary tumor (either by endoscopic ultrasound, operative, or percutaneous approaches) should be avoided because of the high risk of tumor seeding associated with these procedures.

**DONOR CRITERIA: LIVING DONOR LIVER TRANSPLANT**

Donor morbidity and mortality are prime concerns in donors undergoing right lobe, left lobe, or left lateral segment donor partial hepatectomy as part of living donor liver transplantation. Partial hepatectomy is a technically demanding surgery, the success of which may be related to the availability of an experienced surgical team. In 2000, the American Society of Transplant Surgeons proposed the following guidelines for living donors:

- Should be healthy individuals who are carefully evaluated and approved by a multidisciplinary team including hepatologists and surgeons to assure that they can tolerate the procedure
- Should undergo evaluation to assure that they fully understand the procedure and associated risks
- Should be of legal age and have sufficient intellectual ability to understand the procedures and give informed consent
- Should be emotionally related to the recipients
- Must be excluded if the donor is felt or known to be coerced
- Need to have the ability and willingness to comply with long-term follow-up

***Cross-reference:***

**MP 9.013** Isolated Small Bowel Transplant and Small Bowel-Liver and Multivisceral Transplant

**II. PRODUCT VARIATIONS**

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This policy is only applicable to certain programs and products administered by Capital BlueCross and subject to benefit variations as discussed in Section VI. Please see additional information below.

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FEP PPO - Refer to FEP Medical Policy Manual. The FEP Medical Policy manual can be found at: <https://www.fepblue.org/benefit-plans/medical-policies-and-utilization-management-guidelines/medical-policies>

**III. DESCRIPTION/BACKGROUND**

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Solid organ transplantation offers a treatment option for patients with different types of end stage organ failure that can be lifesaving or provide significant improvements to a patient’s quality of life. Many advances have been made in the last several decades to reduce perioperative complications. Available data supports improvement in long-term survival as well as improved quality of life particularly for liver, kidney, pancreas, heart, and lung transplants. Allograft rejection remains a key early and late complication risk for any organ transplantation. Transplant recipients require life-long immunosuppression to prevent rejection. Patients are prioritized for transplant by mortality risk and severity of illness criteria developed by Organ Procurement and Transplantation Network and United Network of Organ Sharing.

**LIVER TRANSPLANTATION**

Liver transplantation is routinely performed as a treatment of last resort for patients with end-stage liver disease. Liver transplantation may be performed with liver donation after a brain or cardiac death or with a liver segment donation from a living donor. Certain populations are prioritized as Status 1A (eg, acute liver failure with a life expectancy of fewer than 7 days without a liver transplant) or Status 1B (pediatric patients with chronic liver disease). Following Status 1, donor livers are prioritized to those with the highest scores on the Model for End-stage Liver Disease (MELD) and Pediatric End-stage Liver Disease (PELD) scales. Due to the scarcity of donor livers, a variety of strategies have been developed to expand the donor pool. For example, a split graft refers to dividing a donor liver into 2 segments that can be used for 2 recipients. Living donor (LD) liver transplantation (LT) is now commonly performed for adults and children from a related or unrelated donor. Depending on the graft size needed for the recipient, either the right lobe, left lobe, or the left lateral segment can be used for LD LT. In addition to addressing the problem of donor organ scarcity, LD LT allows the procedure to be scheduled electively before the recipient’s condition deteriorates or serious complications develop. Living donor LT also shortens the preservation time for the donor liver and decreases disease transmission from donor to recipient.

**REGULATORY STATUS**

Solid organ transplants are a surgical procedure and, as such, are not subject to regulation by the U.S. Food and Drug Administration (FDA).

The FDA regulates human cells and tissues intended for implantation, transplantation, or infusion through the Center for Biologics Evaluation and Research, under Code of Federal Regulation Title 21, parts 1270 and 1271. Solid organs used for transplantation are subject to these regulations.

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**IV. RATIONALE**

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**SUMMARY OF EVIDENCE**

For individuals who have hepatocellular disease who receive a liver transplant, the evidence includes registry studies, and systematic reviews. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. Studies on liver transplantation for viral hepatitis have found that survival is lower than for other liver diseases. Although these statistics raise questions about the most appropriate use of a scarce resource (donor livers), the long-term survival rates are significant in a group of patients who have no other treatment options. Also, survival can be improved by the eradication of the hepatitis virus before transplantation. For patients with NASH, overall survival rates have been shown to be similar to other indications for liver transplantation. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have primary hepatocellular carcinoma who receive a liver transplant, the evidence includes systematic reviews of observational studies. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. In the past, long-term outcomes in patients with primary hepatocellular malignancies had been poor (19%) compared with the overall survival of liver transplant recipients. However, the recent use of standardized patient selection criteria (eg, the Milan criteria diameter) has dramatically improved overall survival rates. In appropriately selected patients, a liver transplant has been shown to result in higher survival rates than resection. In patients who present with unresectable organ-confined disease, transplant represents the only curative approach. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have extrahepatic cholangiocarcinoma who receive a liver transplant, the evidence includes registry studies and a systematic reviews of observational studies. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. For patients with extrahepatic (hilar or perihilar) cholangiocarcinoma who are treated with adjuvant chemotherapy, 5 year survival rates have been reported as high as 76%. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have intrahepatic cholangiocarcinoma who receive a liver transplant, the evidence includes registry studies. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. In a registry study comparing outcomes in patients with intrahepatic cholangiocarcinoma who received liver transplantation to those who received surgical resection of the liver, no differences were found in OS, length of stay, or unplanned 30-day readmission rates between groups. Additional studies reporting survival rates in patients with intrahepatic cholangiocarcinoma or in mixed populations of patients with extrahepatic and intrahepatic cholangiocarcinoma have reported 5-year survival rates of less than 30%. The evidence is insufficient to determine the effects of the technology on health outcomes.

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For individuals who have metastatic neuroendocrine tumors who receive a liver transplant, the evidence includes systematic reviews of case series. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. In select patients with nonresectable, hormonally active liver metastases refractory to medical therapy, liver transplantation has been considered as an option to extend survival and minimize endocrine symptoms. While some centers may perform liver transplants on select patients with neuroendocrine tumors, the available studies are limited by their heterogeneous populations. Further studies are needed to determine the appropriate selection criteria. The evidence is insufficient to determine the effects of the technology on health outcomes.

For individuals who have pediatric hepatoblastoma who receive a liver transplant, the evidence includes case series. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. The literature on liver transplantation for pediatric hepatoblastoma is limited, but case series have demonstrated good outcomes and high rates of long-term survival. Additionally, nonmetastatic pediatric hepatoblastoma is among in United Network for Organ Sharing criteria for patients eligible for liver transplantation. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have a failed liver transplant who receive a liver retransplant, the evidence includes observational studies. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. Case series have demonstrated favorable outcomes with liver retransplantation in certain populations, such as when criteria for an original liver transplantation are met for retransplantation. While some evidence has suggested outcomes after retransplantation may be less favorable than for initial transplantation in some patients, long-term survival benefits have been demonstrated. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals with indications for liver and kidney transplant who receive a CLKT, the evidence includes registry studies. Relevant outcomes include overall survival, morbid events, and treatment-related morbidity and mortality. Most of the evidence involves adults with cirrhosis and kidney failure. Indications for CLKT in children are rare and often congenital and include liver-based metabolic abnormalities affecting the kidney, along with structural diseases affecting both the liver and kidney. In both adults and children, comparisons with either liver or kidney transplantation alone would suggest that CLKT is no worse, and possibly better, for graft and patient survival. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

**V. DEFINITIONS**

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**ALBUMIN** refers to one of a group of simple proteins widely distributed in plant and animal tissues. It is found in the blood as serum albumin, in milk as lactalbumin, and in egg white as ovalbumin.



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**BILIRUBIN** refers to the orange-colored or yellowish pigment in the bile. It is derived from hemoglobin of red blood cells that have completed their life span and are destroyed and ingested by the macrophage system of the liver, spleen, and red bone marrow.

**BLUE QUALITY CENTERS FOR TRANSPLANT (BQCT)** is a cooperative effort of the Blue Cross and Blue Shield Plans, the Blue Cross and Blue Shield Association and participating medical institutions to provide patients who need transplants with access to leading centers through a coordinated, streamlined program of transplant management.

**CADAVER** refers to a dead body or corpse.

**END-STAGE** refers to the final phase of a disease process.

**EXTRAHEPATIC** refers to outside or unrelated to the liver.

**HEPATOCELLULAR** refers to the cells of the liver.

**MACROVASCULAR** refers to the larger blood vessels in the body.

**PROTHROMBIN TIME** is the time it takes for clotting to occur after prothromboplastin and calcium are added to decalcified plasma.

**UNITED NETWORK OF ORGAN SHARING (UNOS)** is an organization established in 1984 to facilitate donation of organs for possible transplantation.

**VI. BENEFIT VARIATIONS**

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The existence of this medical policy does not mean that this service is a covered benefit under the member's health benefit plan. Benefit determinations should be based in all cases on the applicable health benefit plan language. Medical policies do not constitute a description of benefits. A member's health benefit plan governs which services are covered, which are excluded, which are subject to benefit limits and which require preauthorization. There are different benefit plan designs in each product administered by Capital Blue Cross. Members and providers should consult the member's health benefit plan for information or contact Capital Blue Cross for benefit information.

**VII. DISCLAIMER**

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*Capital Blue Cross's medical policies are developed to assist in administering a member's benefits, do not constitute medical advice and are subject to change. Treating providers are solely responsible for medical advice and treatment of members. Members should discuss any medical policy related to their coverage or condition with their provider and consult their benefit information to determine if the service is covered. If there is a discrepancy between this medical policy and a member's benefit information, the benefit information will govern. If a provider or a member has a question concerning the application of this medical policy to a specific member's plan of benefits, please contact Capital Blue Cross' Provider Services or Member*

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*Services. Capital Blue Cross considers the information contained in this medical policy to be proprietary and it may only be disseminated as permitted by law.*

### VIII. CODING INFORMATION

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**Note:** This list of codes may not be all-inclusive, and codes are subject to change at any time. The identification of a code in this section does not denote coverage as coverage is determined by the terms of member benefit information. In addition, not all covered services are eligible for separate reimbursement.

#### Covered when medically necessary:

Procedure Codes								
47133	47135	47140	47141	47142	47143	47144	47145	47146
47147	47399	S2152						

ICD-10-CM Diagnosis Code	Description
B15.0	Hepatitis A with hepatic coma
B15.9	Hepatitis A without hepatic coma
B16.0	Acute hepatitis B with delta-agent with hepatic coma
B16.1	Acute hepatitis B with delta-agent without hepatic coma
B16.2	Acute hepatitis B without delta-agent with hepatic coma
B16.9	Acute hepatitis B without delta-agent and without hepatic coma
B17.0	Acute delta-(super) infection of hepatitis B carrier
B17.10	Acute hepatitis C without hepatic coma
B17.11	Acute hepatitis C with hepatic coma
B17.2	Acute hepatitis E
B17.8	Other specified acute viral hepatitis
B18.0	Chronic viral hepatitis B with delta-agent
B18.1	Chronic viral hepatitis B without delta-agent
B18.2	Chronic viral hepatitis C
B18.8	Other chronic viral hepatitis
B19.10	Unspecified viral hepatitis B without hepatic coma
B19.11	Unspecified viral hepatitis B with hepatic coma
B19.20	Unspecified viral hepatitis C without hepatic coma
B19.21	Unspecified viral hepatitis C with hepatic coma
B19.9	Unspecified viral hepatitis without hepatic coma
B25.1	Cytomegaloviral hepatitis
B66.1	Clonorchiasis
B66.5	Fasciolopsiasis
C22.0	Liver cell carcinoma

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<b>ICD-10-CM Diagnosis Code</b>	<b>Description</b>
C22.1	Intrahepatic bile duct carcinoma
C22.2	Hepatoblastoma
C22.3	Angiosarcoma of liver
C22.4	Other sarcomas of liver
D13.4	Benign neoplasm of liver
D13.5	Benign neoplasm of extrahepatic bile ducts
D64.1	Secondary sideroblastic anemia due to disease
D64.2	Secondary sideroblastic anemia due to drugs and toxins
D64.3	Other sideroblastic anemias
D64.4	Congenital dyserythropoietic anemia
D81.810	Biotinidase deficiency
D84.1	Defects in the complement system
E70.0	Classical Phenylketonuria
E70.1	Other hyperphenylalaninemias
E70.5	Disorders of Tryptophan metabolism
E70.81	aromatic l-amino acid decarboxylase deficiency
E70.89	Other disorders of aromatic amino- acid metabolism
E71.42	Carnitine deficiency due to inborn errors of metabolism
E72.00	Disorders of amino-acid transport, unspecified
E72.01	Cystinuria
E72.02	Hartnup's disease
E72.03	Lowe's syndrome
E72.04	Cystinosis
E72.09	Other disorders of amino-acid transport
E74.00	Glycogen storage disease, unspecified
E74.01	von Gierke disease
E74.02	Pompe disease
E74.03	Cori disease
E74.04	McArdle disease
E74.05	Lysosome-associated membrane protein 2 [LAMP2] deficiency
E74.09	Other glycogen storage disease
E74.10	Disorder of fructose metabolism, unspecified
E74.11	Essential fructosuria
E74.12	Hereditary fructose intolerance
E74.19	Other disorders of fructose metabolism
E74.20	Disorders of galactose metabolism, unspecified

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<b>ICD-10-CM Diagnosis Code</b>	<b>Description</b>
E74.21	Galactosemia
E74.29	Other disorders of galactose metabolism
E74.31	Sucrase-isomaltase deficiency
E74.39	Other disorders of intestinal carbohydrate absorption
E74.4	Disorders of pyruvate metabolism and gluconeogenesis
E70.81	Aromatic l-amino acid decarboxylase deficiency
E70.89	Other disorders of aromatic amino-acid metabolism
E74.81	Disorders of glucose transport, not elsewhere classified
E74.810	Glucose transporter protein type 1 deficiency
E74.818	Other disorders of glucose transport
E74.819	Disorders of glucose transport, unspecified
E74.89	Other specified disorders of carbohydrate metabolism
E74.9	Disorder of carbohydrate metabolism, unspecified
E78.0	Pure hypercholesterolemia
E78.1	Pure hyperglyceridemia
E78.2	Mixed hyperlipidemia
E78.3	Hyperchylomicronemia
E78.41	Elevated Lipoprotein(a)
E78.49	Other hyperlipidemia
E78.5	Hyperlipidemia, unspecified
E78.6	Lipoprotein deficiency
E78.70	Disorder of bile acid and cholesterol metabolism, unspecified
E78.71	Barth syndrome
E78.72	Smith-Lemli-Opitz syndrome
E78.79	Other disorders of bile acid and cholesterol metabolism
E78.81	Lipoid dermatoarthritis
E78.89	Other lipoprotein metabolism disorders
E78.9	Disorder of lipoprotein metabolism, unspecified
E80.0	Hereditary erythropoietic porphyria
E80.1	Porphyria cutanea tarda
E80.20	Unspecified porphyria
E80.21	Acute intermittent (hepatic) porphyria
E80.29	Other porphyria
E80.3	Defects of catalase and peroxidase
E80.4	Gilbert syndrome
E80.5	Crigler-Najjar syndrome

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<b>ICD-10-CM Diagnosis Code</b>	<b>Description</b>
E80.6	Other disorders of bilirubin metabolism
E80.7	Disorder of bilirubin metabolism, unspecified
E83.00	Disorder of copper metabolism, unspecified
E83.01	Wilson's disease
E83.09	Other disorders of copper metabolism
E83.10	Disorder of iron metabolism, unspecified
E83.11	Hemochromatosis due to repeated red blood cell transfusions
E83.110	Hereditary hemochromatosis
E83.111	Hemochromatosis due to repeated red blood cell transfusions
E83.118	Other hemochromatosis
E83.119	Hemochromatosis, unspecified
E83.19	Other disorders of iron metabolism
E85.0	Non-neuropathic hereditary amyloidosis
E85.1	Neuropathic hereditary amyloidosis
E85.2	Hereditary amyloidosis, unspecified
E85.3	Secondary systemic amyloidosis
E85.4	Organ-limited amyloidosis
E85.81	Light chain (AL) amyloidosis
E85.82	Wild-type transthyretin-related (ATTR) amyloidosis
E85.89	Other amyloidosis
E88.01	Alpha-1-antitrypsin deficiency
E88.9	Metabolic disorder, unspecified
G60.0	Hereditary motor and sensory neuropathy
G60.1	Refsum's disease
G60.2	Neuropathy in association with hereditary ataxia
G60.3	Idiopathic progressive neuropathy
G60.8	Other hereditary and idiopathic neuropathies
I74.8	Embolism and thrombosis of other arteries
I82.0	Budd-Chiari syndrome
K70.30	Alcoholic cirrhosis of liver without ascites
K70.31	Alcoholic cirrhosis of liver with ascites
K71.0	Toxic liver disease with cholestasis
K71.10	Toxic liver disease with hepatic necrosis, without coma
K71.11	Toxic liver disease with hepatic necrosis, with coma
K71.2	Toxic liver disease with acute hepatitis
K71.3	Toxic liver disease with chronic persistent hepatitis

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<b>ICD-10-CM Diagnosis Code</b>	<b>Description</b>
K71.4	Toxic liver disease with chronic lobular hepatitis
K71.5	Toxic liver disease with chronic active hepatitis
K71.50	Toxic liver disease with chronic active hepatitis without ascites
K71.51	Toxic liver disease with chronic active hepatitis with ascites
K71.6	Toxic liver disease with hepatitis, not elsewhere classified
K71.7	Toxic liver disease with fibrosis and cirrhosis of liver
K71.8	Toxic liver disease with other disorders of liver
K71.9	Toxic liver disease; Toxic liver disease, unspecified
K74.1	Hepatic sclerosis
K74.2	Hepatic fibrosis with hepatic sclerosis
K74.3	Primary biliary cirrhosis
K74.4	Secondary biliary cirrhosis
K74.5	Biliary cirrhosis, unspecified
K74.60	Unspecified cirrhosis of liver
K74.69	Other cirrhosis of liver
K75.4	Autoimmune hepatitis
K75.81	Nonalcoholic steatohepatitis (NASH)
K77	Liver disorders in diseases classified elsewhere (code first underlying disease)
K83.01	Primary sclerosing cholangitis
K83.1	Obstruction of bile duct
M34.83	Systemic sclerosis with polyneuropathy
Q44.2	Atresia of bile ducts
Q44.6	Cystic disease of liver
S36.112A	Contusion of liver, initial encounter
S36.112D	Contusion of liver, subsequent encounter
S36.114A	Minor laceration of liver, initial encounter
S36.114D	Minor laceration of liver, subsequent encounter
S36.115A	Moderate laceration of liver, initial encounter
S36.115D	Moderate laceration of liver, subsequent encounter
S36.116A	Major laceration of liver, initial encounter
S36.116D	Major laceration of liver, subsequent encounter
S36.113A	Laceration of liver, unspecified degree, initial encounter
S36.113D	Laceration of liver, unspecified degree, subsequent encounter
S36.113S	Laceration of liver, unspecified degree, sequela
S36.118A	Other injury of liver, initial encounter

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<b>ICD-10-CM Diagnosis Code</b>	<b>Description</b>
S36.118D	Other injury of liver, subsequent encounter
T86.41	Liver transplant rejection
T86.42	Liver transplant failure
T86.49	Other complications of liver transplant
Z52.6	Liver donor

### IX. REFERENCES

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**X. POLICY HISTORY**

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<b>MP 9.006</b>	<b>2/11/20 Consensus review.</b> No changes to policy statements. References Updated.
	<b>10/1/20 Administrative update.</b> Added new codes and removed end-dated codes effective 10-1-20.

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<b>12/1/2021 Consensus review.</b> No change to policy statement. References reviewed and updated.
<b>12/30/2022 Consensus review.</b> No change to policy statement. References, rationale and background reviewed and updated.
<b>8/30/2023 Administrative review.</b> New diagnosis code E74.05 added to policy from new code update. Effective date 10/1/2023.
<b>8/31/2023 Consensus review.</b> No change to policy statement. Rationale updated. References reviewed and updated. Coding reviewed.
<b>1/19/2024 Administrative update.</b> Clinical benefit added.

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