

APPENDIX A

Patient Selection Criteria

I. Heart patient selection criteria

A. Indications

1. Irremediable terminal cardiac disease with severely compromised survival despite all other available medical and surgical therapies with survival estimates based on standard heart failure risk assessments.
2. Any patient who is continually on the waiting list for more than two years will require verification from the transplant team that he or she is still a candidate for transplantation and continues to meet the patient selection criteria as described below.
3. Age seventy years or younger (older patients will be considered on a case-by-case basis).
4. Patient selection criteria as published in the September 2006 issue of the "Journal of Heart and Lung Transplantation," Volume 25, pages 1024 through 1042.

B. Relative contraindications

Body mass index greater than thirty-five kilograms per meters squared, or percent body mass index of greater than one hundred and forty percent.

C. Relative contraindications for pediatric candidates (to be reviewed on a case by case basis):

1. Pre-maturity (less than thirty-six weeks gestation);
2. Small size (less than two to two and five tenths kilograms);
3. Cardiac anatomic abnormalities:
 - a. Significant pulmonary artery hypoplasia
 - b. Uncorrectable pulmonary venous abnormalities
 - c. Ectopia cordis
4. Cardiac physiologic abnormalities:

Elevated pulmonary vascular resistance that is irreversible with pulmonary vasodilator testing (greater than six Wood units per meter squared);

5. Impaired and/or irreversible dysfunction or abnormality of other organ systems, such as acute renal failure, acute hepatic failure, multisystem organ failure, severe dysmorphology or genetic abnormality, profound metabolic or neuromuscular / neurologic / central nervous system disorder;
6. Severe or significant systemic illness such as active infection (pneumonitis, septicemia, fever of unknown origin) human immunodeficiency virus infection, hepatitis B and possible hepatitis C infection, active malignancy;
7. Psychosocial pathology, including alcohol or substance abuse in parents or patients, child abuse or neglect, psychiatric illness in patient, strong and irremediable history of non-compliance;
8. Parental non-compliance with physician recommendations for pediatric candidates to have all age appropriate immunizations.

D. Contraindications

1. Significant active infection, unless related to a left ventricular assist device or uncontrolled malignancy;
2. Diabetes with end organ damage other than non-proliferative retinopathy or poor glycemic control despite optimal effort;
3. Pulmonary artery hypertension and elevated pulmonary vascular resistance (PVR) when PVR is greater than five Woods units, the PVR index is greater than six, or the transpulmonary gradient exceeds sixteen to twenty millimeters of mercury;
4. Clinically severe symptomatic cerebrovascular disease that is not amendable to revascularization;
5. Alcohol or substance dependency not consistent with screening criteria in Appendix B of this rule;
6. Absence of adequate external psychosocial support;
7. Other life-limiting illnesses;
8. Irreversible hepatic or renal dysfunction, unless the patient is being considered for multiple organ transplant;
9. Non-compliance.

II. Heart-lung patient selection criteria

- A. General indications: All patients considered for heart-lung transplantation shall:

1. Be less than sixty years of age;
 2. Be free from other major organ system failure;
 3. Have an expected survival time of not more than twelve to twenty-four months without a transplant.
- B. Specific indications: patients with the following disease processes are considered as candidates for transplantations. They should be functionally limited, but not totally disabled. There is a “transplant window” during which the patient has a gradual decline from the natural history of his or her disease, but has not deteriorated so much that he or she is no longer a viable transplant candidate. Patients to be considered include, but are not limited to, those with the following diagnoses:
1. Eisenmenger’s syndrome;
 2. Primary pulmonary hypertension;
 3. Cardiomyopathy with pulmonary hypertension;
 4. Emphysema;
 5. Alpha-1 antitrypsin deficiency;
 6. Pulmonary fibrosis;
 7. Cystic fibrosis;
 8. Bronchiectasis;
 9. Bronchopulmonary dysplasia;
 10. Post-transplant obliterative bronchiolitis;
 11. Pulmonary disease in young infant for technical surgical issues (typically due to the airway size).
- C. Contraindications
1. Absolute:
 - a. Significant systemic or multi-system disease;
 - b. Active or systemic infection limiting survival;
 - c. Significant hepatic disease;
 - d. Significant renal disease;

- e. Current cigarette smoking (minimum of six months abstinence and a commitment to continue non-smoking post-transplant);
- f. Psychiatric illness.

2. Potential:

- a. Previous cardiac or thoracic surgery;
- b. Diabetes mellitus;
- c. Peptic ulcer disease;
- d. Alcohol or substance dependency not consistent with screening criteria in Appendix B of this rule;
- e. History of cancer;
- f. Cachexia (less than seventeen body mass index) or obesity (greater than thirty body mass index);
- g. Non-compliance;
- h. Corticosteroid therapy (greater than twenty milligrams per day).

III. Single/double lung patient selection criteria

A. General indications: All patients considered for pulmonary transplantation shall be suffering from end-stage pulmonary disease and shall:

- 1. Be less than sixty-five years of age; (patients older than sixty-five will be reviewed on a case by case basis)
- 2. Be free from other major organ failure;
- 3. Be free from major psychosocial problems;
- 4. Be capable of participating in pre- and post-operative rehabilitation programs and follow-up;
- 5. Have an expected survival time of less than eighteen months without a transplant.

B. Specific indications: Patients with the following disease processes will be considered for lung transplantation:

- 1. Eisenmenger's physiology, chronic obstructive pulmonary disease (COPD), emphysema, Alpha-1 antitrypsin deficiency;
- 2. Primary pulmonary hypertension;

3. Pulmonary fibrosis (primary or secondary);
4. Cystic fibrosis;
5. Bronchiectasis;
6. Other, including but not limited to, sarcoidosis, systemic lupus, pulmonary hemosiderosis, Bronchiolitis obliterans;
7. Advanced pediatric lung disease.

C. Contraindications

1. Absolute:

- a. Significant systemic or multi-system disease;
- b. Active or systemic infection limiting survival;
- c. Current cigarette smoking (minimum of six months abstinence and a commitment to continue non-smoking post-transplant);
- d. Malignancy in the last two years, with the exception of cutaneous squamous and basal cell tumors. In general a five year disease free interval is prudent;
- e. Major psychiatric illness;

2. Potential:

- a. Previous cardiac or thoracic surgery;
- b. Diabetes mellitus;
- c. Peptic ulcer disease;
- d. Alcohol or substance dependency not consistent with screening criteria in Appendix B of this rule;
- e. Cachexia (less than seventeen body mass index) or obesity (greater than thirty body mass index);
- f. Corticosteroid therapy (greater than twenty milligrams per day);
- g. Psychosocial issues;
- h. Non-compliance.

IV. Hepatic patient selection criteria

- A. General indications: In general, liver transplantation is indicated in children and adults suffering from irreversible liver dysfunction or the effects of liver dysfunction after alternative medical and surgical treatments have been utilized and where the benefits of transplantation outweigh the risk of alternative modalities.
- B. Specific indications
1. Acute hepatic fulminant failure;
 2. Extra-hepatic biliary atresia or hypoplasia;
 3. Inborn errors of metabolism:
 - a. Alpha I antitrypsin deficiency;
 - b. Crigler-Najjar disease, Type I;
 - c. Byler's disease;
 - d. Glycogen storage disease (O and IV);
 - e. Wilson's disease;
 - f. Hemochromatosis;
 - g. Tyrosinemia;
 - h. Wolman's disease;
 - i. Familial amyloidotic polyneuropathy (FAP);
 - j. Primary hyperoxaluria, Type I;
 - k. Other.
 4. Sclerosing cholangitis;
 5. Hepatic vein thrombosis (Budd-Chiari);
 6. Hepatocellular Carcinoma (HCC), Stage I or II, or University of California at San Francisco (UCSF) /TNM. Criteria.
 7. Cirrhosis:
 - a. Alcoholic cirrhosis (see alcohol or substance dependence screening criteria in Appendix B of this rule);

- b. Biliary cirrhosis (primary or secondary): Caroli, choledochal cyst, congenital cholestasis [progressive familial intra-hepatic cholestasis (PFIC)], iatrogenic biliary tree injury/damage, trauma;
 - c. Chronic active hepatitis (A, B, C, non A, non B, autoimmune);
 - d. Congenital biliary cirrhosis;
 - e. Cryptogenic cirrhosis;
 - f. Cystic fibrosis;
 - g. Hemochromatosis;
 - h. Alpha I Antitrypsin Deficiency;
 - i. Non-alcoholic steatosis hepatitis (NASH);
 - j. Viral cirrhosis;
 - k. Other.
- 8. Congenital hepatic fibrosis;
 - 9. Controlled biliary sepsis resulting from acute (or chronic) hepatic artery thrombosis (ischemic coagulopathy);
 - 10. Hepato-pulmonary syndrome, with cirrhosis;
 - 11. Polycystic liver disease with symptoms such as: portal hypertension, Budd-Chiari-like symptoms, refractory and unmanageable ascites following cyst fenestration;
 - 12. Porto-pulmonary hypertension in the presence of cirrhosis and mean pulmonary artery pressures of greater than thirty-five (35) millimeters of mercury;

C. Indications for re-transplantation

- 1. Primary non-function;
- 2. Irreversible vascular compromise of either the hepatic artery, portal vein, or hepatic vein;
- 3. Recurrent primary disease;
- 4. Intractable, acute, or chronic rejection;
- 5. Biliary disease not correctable by any mechanism other than transplantation;

6. Small for size syndrome;
 7. Poor early graft function (PEGF);
- D. Indications for special cases to be reviewed on a case-by-case basis
1. Stage III tumors outside of the University of UCSF/TNM criteria;
 2. Slow-growing metastatic leiomyosarcoma and neuroendocrine tumors, including metastatic carcinoid syndrome and hemangioendotheliomas;
 3. Cholangiocarcinoma transplanted under approved institutional protocol;
 4. Hepatitis C (re-transplant within one year of recurrent disease);
- E. Contraindications
1. Alcohol or substance dependency not consistent with screening criteria in Appendix B of this rule;
 2. Active infection outside hepatobiliary system limiting survival;
 3. Significant cardiac, pulmonary, or nervous system failure (this does not apply for patients being considered for heart and/or lung transplant);
 4. Unstable current psychotic disease (pre-liver failure);
 5. Uncontrolled malignancy;
 6. Acquired immune deficiency syndrome (AIDS);
 7. Persistent pattern of non-compliance considered likely to interfere with following a disciplined medical regime;
- V. Small bowel: Intestinal / small bowel and liver patient selection criteria
- A. Background: Small intestinal transplantation, combined liver/intestinal transplantation, and multivisceral transplantation are operations that are reserved for life-threatening conditions, but no longer considered investigational. When reserved for patients with no possibility of long-term survival without transplantation, the procedure should be strongly considered. Factors governing success of the procedure will include the general health of the recipient before transplant, the size of the center, the experience of the physicians within that center with small bowel or liver transplantation and/or related procedures, and recent advances in surgical techniques, graft monitoring techniques, immunosuppressive regimens, and antiviral therapies.
- B. General indications: Isolated small intestinal transplantation is an operation reserved for patients with irreversible short bowel syndrome (or intestinal

failure despite intact intestinal length) associated with life-threatening complications due to either parenteral nutrition or the underlying disorders. When prospects for weaning from parenteral nutrition are nil, and patients have experienced multiple serious episodes of septicemia, progressive loss of vascular access are at risk for serious morbidity and mortality from the disease itself, or are at risk for progressive cholestasis associated with parenteral nutrition, isolated small intestinal transplantation may be considered.

When candidates have suffered irreversible liver damage (usually associated with prolonged parenteral nutrition) in addition to irreversible intestinal failure, liver / small intestinal transplantation should be considered. If the candidate's disease process is one that renders gastroduodenal function unacceptable, or when localized tumors or other causes of vascular occlusion seriously compromise the arterial blood supply to the stomach, liver, small bowel, and pancreas, multivisceral transplantation may be performed.

C. Specific indications

1. Surgical short bowel syndrome due to:
 - a. Volvulus;
 - b. Gastroschisis;
 - c. Necrotizing enterocolitis;
 - d. Hirschsprung's disease;
 - e. Congenital atresias;
 - f. Crohn's disease;
 - g. Trauma;
 - h. Mesenteric vascular insufficiency;
 - i. Localized intra-abdominal tumors such as desmoid tumors or inflammatory pseudotumor;
 - j. Other causes of surgical short bowel syndrome;
2. Secretory diarrhea associated with uncorrectable malabsorption:
 - a. Microvillus inclusion disease;
 - b. Tufting enteropathy;
 - c. Intestinal pseudo-obstruction;

- d. Other life-threatening diarrheal disorders uncorrectable by medical or surgical means short of transplantation.
- 3. Liver failure in association with intestinal failure:
 - a. Total parenteral nutrition (TPN) -associated liver failure;
 - b. Other causes of liver failure (such as primary sclerosing cholangitis or biliary atresia) in association with intestinal failure.
- 4. Other Consortium approved indications for transplantation.

D. Contraindications

- 1. Alcohol or drug dependence not consistent with screening criteria in Appendix B of this rule;
- 2. Active infection outside the hepatobiliary system limiting survival;
- 3. Disseminated, non-resectable malignancy;
- 4. Insufficient venous patency to guarantee central venous access;
- 5. Severe dysfunction of other organ systems (cardiac, pulmonary, vascular, renal, neurologic) rendering transplantation risk unacceptable;
- 6. Other life-threatening, uncorrectable illnesses not referable to the gastrointestinal system;
- 7. Unstable, uncontrollable psychiatric illness;
- 8. Proven non-compliance.

VI. Pancreas patient selection criteria

A. Indications

- 1. Group I: Insulin dependent diabetes mellitus Type I (juvenile – onset diabetes mellitus or JODM) with secondary complications but no renal or early involvement. Early pancreatic transplantation may be indicated for those who cannot be adequately controlled with insulin and/or with chronic complications of diabetes likely to be impacted by a pancreas transplant.
- 2. Group II: Insulin dependent diabetes mellitus Type I with secondary complications and renal involvement in absence of morbid obesity. Synchronous pancreatic and renal transplantation may be indicated.

3. Group III: Insulin dependent diabetes mellitus Type I or Type II with secondary complications, end-stage renal disease and previous renal transplant. Asynchronous pancreatic transplantation may be indicated.

B. Contraindications

1. Alcohol or substance dependency not consistent with screening criteria in Appendix B of this rule;
2. Active sepsis;
3. End-stage organ dysfunction:
 - a. Pulmonary;
 - b. Cardiac;
 - c. Cerebrovascular;
 - d. Hematological;
4. Inability to understand the transplant procedure and care for self after receiving a new organ;
5. Uncontrolled malignancy.

C. Relative contraindications

1. Body mass index greater than thirty-five;
2. Non-compliance.

VII. Pancreas islet patient selection criteria

A. Indications

1. All recipients must have Type I insulin-dependent diabetes mellitus with absent C-Peptide;
2. Previous kidney transplant [or kidney-pancreas transplant with failed pancreas with satisfactory kidney graft function];
3. Glomerular filtration rate (GFR) greater than fifty based on calculated GFR or based on a twenty-four hour urine study or iothalamate clearance study, as appropriate. This is to only apply to patients who have not previously received a kidney transplant and are potential kidney recipients;

4. Kidney transplant candidates with Type I insulin-dependent diabetes mellitus.

B. Contraindications

1. History of malignancy (except adequately treated localized basal cell carcinoma of skin without evidence of recurrence, or other cancers considered cured by therapy);
2. Patients with current episodes of acute renal allograft rejection;
3. Patients with panel reactive antibody (PRA) level greater than fifty percent;
4. Alcohol or substance dependency not consistent with screening criteria in Appendix B of this rule;
5. Active infection;
6. Non-compliance.