

Name of Policy:

Treatment of Twin-Twin Transfusion Syndrome with Amnioreduction and/or Fetoscopic Laser Therapy

Policy #: 072 Latest Review Date: November 2010 Category: Surgery Policy Grade: Active Policy but no longer

scheduled for regular literature reviews and

updates.

Background/Definitions:

As a general rule, benefits are payable under Blue Cross and Blue Shield of Alabama health plans only in cases of medical necessity and only if services or supplies are not investigational, provided the customer group contracts have such coverage.

The following Association Technology Evaluation Criteria must be met for a service/supply to be considered for coverage:

- 1. The technology must have final approval from the appropriate government regulatory bodies;
- 2. The scientific evidence must permit conclusions concerning the effect of the technology on health outcomes;
- 3. The technology must improve the net health outcome;
- 4. The technology must be as beneficial as any established alternatives;
- 5. The improvement must be attainable outside the investigational setting.

Medical Necessity means that health care services (e.g., procedures, treatments, supplies, devices, equipment, facilities or drugs) that a physician, exercising prudent clinical judgment, would provide to a patient for the purpose of preventing, evaluating, diagnosing or treating an illness, injury or disease or its symptoms, and that are:

- 1. In accordance with generally accepted standards of medical practice; and
- 2. Clinically appropriate in terms of type, frequency, extent, site and duration and considered effective for the patient's illness, injury or disease; and
- 3. Not primarily for the convenience of the patient, physician or other health care provider;
- 4. Not more costly than an alternative service or sequence of services at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of that patient's illness, injury or disease.

Description of Procedure or Service:

Twin-twin transfusion syndrome (TTTS) is a severe complication of monozygotic twinning. It is relatively common, occurring in 10% - 15% of all monochorionic twins. It is often a lethal condition, accounting for 17% of perinatal deaths in twins overall. Without treatment, perinatal mortality exceeds 80%.

Twin-Twin Transfusion Syndrome is diagnosed in the presence of:

- 1. Monochorionic-diamniotic twin pregnancy with discordance in size.
- 2. Polyhydramnios in the gestational sac of the recipient twin.
- 3. Oligohydramnios in the gestational sac of the donor twin.
- 4. Polyuria of the recipient.
- 5. Oliguria of the donor.

Two techniques have recently become the standard of treatment for this condition:

- 1. Amnioreduction—Using ultrasound guidance, a spinal needle is used to remove excess amniotic fluid from the recipient twin (with polyhydramnios).
- 2. Laser coagulation therapy—With local anesthesia and ultrasound guidance, fetoscopic laser is used to coagulate placental vascular anastomoses. This may be preceded by either angiography or Doppler ultrasound to target the vessels for laser therapy. This treatment is frequently performed in centers, which specialize in multiple pregnancies.

Policy:

Amnioreduction (CPT 59001) **meets** Blue Cross and Blue Shield of Alabama's medical criteria for coverage as a treatment of Twin-Twin Transfusion Syndrome.

Fetoscopic laser coagulation therapy (CPT S2411) **meets** Blue Cross and Blue Shield of Alabama's medical criteria for coverage as a treatment of Twin-Twin Transfusion Syndrome.

Blue Cross and Blue Shield of Alabama does not approve or deny procedures, services, testing, or equipment for our members. Our decisions concern coverage only. The decision of whether or not to have a certain test, treatment or procedure is one made between the physician and his/her patient. Blue Cross and Blue Shield of Alabama administers benefits based on the members' contract and corporate medical policies. Physicians should always exercise their best medical judgment in providing the care they feel is most appropriate for their patients. Needed care should not be delayed or refused because of a coverage determination.

Key Points:

The cause and pathophysiology of twin-twin transfusion syndrome remain poorly understood. Classic surgical pathologic studies showed that placental anastomoses could be either superficial (arterioarterial or venovenous) or deep arteriovenous. Superficial anastomoses may have a bidirectional blood flow and deep anastomoses establish a unidirectional flow of blood between the fetuses. The primary defect is thought to be abnormal placentation, with a paucity of superficial anastomoses and the deep anastomoses establish a unidirectional flow of blood from the donor twin to the recipient twin. The donor twin progressively becomes anemic, hypotensive, and hypovolemic, with resultant oligohydramnios and lagging growth; the

recipient twin becomes polycythemic, hypertensive, and hypervolemic, with resultant polyhydramnios, cardiomegaly, and congestive heart failure. Prenatal management strategies are aimed at amelioration of polyhydramnios and/or at correction of the underlying vascular anomalies in the shared placenta.

The optimal treatment in pregnancies that are complicated by TTTS is controversial. Most therapies have evolved only during the past 5 to 10 years and include expectant management, medical therapy (i.e., digoxin), and delivery of the compromised twin, selective feticide, or septostomy. Outcomes from these options have been disappointing. Two treatments, serial amnioreduction and fetoscopic laser ablation of anastomotic vessels, are currently undergoing active investigation. Amnioreduction is the removal of excess amniotic fluid from the recipient's sac to restore normal fluid volume, and reduce the risk of preterm labor and spontaneous rupture of membranes. Fetoscopic laser therapy is designed to correct the underlying abnormality by separating the two fetal circulations. Refinements of laser therapy have focused on the selective ablation of those few arteriovenous anastomoses causing disease. Specific anastomoses can be targeted using angiography, doppler ultrasonography, or direct fetoscopic visualization.

Although there are 3 randomized controlled trials currently in progress, no results have been reported from any of these trials. For example, 1 prospective randomized clinical trial sponsored by the National Institutes of Health (NIH) is comparing aggressive serial amnioreduction with selective fetoscopic laser photocoagulation. The primary evidence supporting amnioreduction with or without laser therapy consists of 15 uncontrolled case series that report consistent evidence of a survival advantage. There was an overall fetal survival Studies conducted since the prior TEC Assessment report only on case series with outcomes similar to those reported in the TEC Assessment. However, complicating the assessment of these procedures is the improved outcomes of those managed conservatively. In a meta-analysis by Skupski et al, there were no differences in outcomes between those treated by either amnioreduction or laser therapy and those treated conservatively. However, small numbers and lack of control for confounding variables do not allow for firm conclusions. For all surviving twins, morbidity remains high, dominated by neurologic cardiovascular and renal complications. For example, the incidence of cerebral palsy and global developmental delay in the surviving twins varies from 4% to 23%.

A November 2006 literature review has found no new information that would alter the current coverage statement for this policy.

November 2008 Update

A literature search has identified no new information that alters the coverage statement of this policy.

Key Words:

TTTS = Twin-twin transfusion syndrome, amnioreduction, laser coagulation therapy, and fetoscopic laser therapy

Approved by Governing Bodies:

Not applicable

Benefit Application:

Coverage is subject to member's specific benefits. Group specific policy will supersede this policy when applicable.

ITS: Home Policy provisions apply

FEP contracts: Special benefit consideration may apply. Refer to member's benefit plan.

Pre-certification/Pre-determination requirements: Not applicable

Coding:

CPT codes: 59001 Amniocentesis, therapeutic amniotic fluid reduction (includes

ultrasound guidance)

S2411 Fetoscopic laser therapy for treatment of twin-to-twin transfusion

syndrome

References:

- 1. Bermudez, Carlos, Becerra, Carlos H., et al. *Placental types and twin-twin transfusion syndrome*. American Journal of Obstetrics and Gynecology, August 2002, Vol. 187, No. 2.
- 2. Crombleholme TM. *The treatment of twin-twin transfusions syndrome*. Semin Pediatr Surg 2003;12(3):175-81.
- 3. Dickinson, Jan E. and Evans, Sharon F. *Obstetric and perinatal outcomes from The Australian and New Zealand Twin-Twin Transfusion Syndrome Registry*. American Journal of Obstetrics and Gynecology, March 2000, Vol. 182, No. 3.
- 4. Hecher, Kurt, Plath, Hanno, et al. *Endoscopic laser surgery versus serial amniocenteses in the treatment of severe twin-twin transfusion syndrome*. American Journal of Obstetrics and Gynecology, March 1999, Vol. 180, No. 3.
- 5. Johnson, Jeffrey R., et al. *Amnioreduction versus septostomy in twin-twin transfusion syndrome*, American Journal of Obstetrics and Gynecology, November 2001, Vol. 185, No. 5.
- 6. Lopriore, E., Nagel, H.T., Vandenbussche, F.P., et al. *Long-term neurodevelopment outcomes in twin-to-twin transfusion syndrome*. Am J Obstet Gynecol 2003;189(5):1314-9.
- 7. Mari, Giancarlo, Detti, Laura, et al. *Long-term outcome in twin-twin transfusion syndrome treated with serial aggressive amnioreduction*, American Journal of Obstetrics and Gynecology, July 2000, Vol. 183, No. 1.
- 8. Mari, Giancarly, Roberts, Alistair, et al. *Perinatal morbidity and mortality rates in severe twin-twin transfusion syndrome: Results of the International Amnioreduction Registry*, American Journal of Obstetrics and Gynecology, September 2001, Vol. 185, No. 3.

- 9. Skupski DW, Gurushanthaiah K, Chasen S. *The effect of treatment of twin-twin transfusion syndrome on the diagnosis-to-delivery interval*. Twin Res 2002; 5(1):1-4.
- 10. Taylor, Myles J.O., Denbow, Mark L., et al. *Antenatal factors at diagnosis that predict outcome in twin-twin transfusion syndrome*. American Journal of Obstetrics and Gynecology, October 2000, Vol. 183, No. 4.
- 11. Wee, L.Y., Fisk, N.M. *The twin-twin transfusion syndrome*. Semin Neonatol 2002; 7(3):187-202.

Policy History:

Medical Policy Group, October 2002

Medical Policy Administration Committee, October 2002

Available for comment December 19, 2002-January 3, 2003

Medical Policy Group, November 2004 (4)

Medical Policy Group, November 2006 (1)

Medical Policy Group, November 2008 (1)

Medical Policy Group, November 2010 (1)

Medical Policy Administration Committee, December 2010

<u>Medical Policy Group, December 2010: Effective December 2, 2010 this is an active Policy but is no longer scheduled for regular literature reviews and updates.</u>

This medical policy is not an authorization, certification, explanation of benefits, or a contract. Eligibility and benefits are determined on a case-by-case basis according to the terms of the member's plan in effect as of the date services are rendered. All medical policies are based on (i) research of current medical literature and (ii) review of common medical practices in the treatment and diagnosis of disease as of the date hereof. Physicians and other providers are solely responsible for all aspects of medical care and treatment, including the type, quality, and levels of care and treatment.

This policy is intended to be used for adjudication of claims (including pre-admission certification, pre-determinations, and pre-procedure review) in Blue Cross and Blue Shield's administration of plan contracts.