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Manual of
High Risk Fetus

Giving Light to New Life

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CHAPTER

Surgical Fetal Therapy: Shunts and Lasers

Archita Shaleen

INTRODUCTION

Fetal surgery is a relatively new field of medicine. Earlier the only treatment of congenital anomalies was planned delivery at tertiary care center and postnatal management. With the availability of better diagnostic techniques and superior understanding of fetal pathophysiology, great success has been achieved in the treatment of previous devastating anomalies.

Fetal surgery is confined to anatomical problems for which correction *in utero* is feasible and may be of significant benefit in the future development and survival of the fetus without involving major risk to the mother.

There are various approaches to fetal intervention, all of which have been developed in the last few decades **(Table 1)**.

USE OF SHUNTS AND LASERS IN FETAL SURGERY

In fetal surgery by shunts and lasers, fetus is manipulated without incision in the uterus. The procedure is done in local or regional anesthesia with or without sedation. Shunt placement is done under sonographic guidance. Laser therapy can be fetoscopic or interstitial laser which is done under ultrasound guidance. They are done by either direct insertion of large bore needle in mother's abdomen or by a small incision. Though the procedure is minimally invasive, there is risk of preterm labor, leaking or bleeding per vaginum. Hence, postprocedure strict monitoring is advised.

FETAL SHUNT SURGERY

A shunt is a hollow tube which is inserted, under ultrasound guidance, through the mother's abdomen and uterus and into the fetus connecting a fluid filled cavity to the amniotic cavity to provide continuous drainage. There are various indications of shunt placement which are described as follows.

Thoracoamniotic Shunt

Fetal thoracic lesions such as pleural effusion (FPE), macrocystic congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration

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Table 1: Various approaches of fetal intervention and its common applications.

Types of surgery	*Common applications*
Open surgery	• Congenital pulmonary airway malformation (CPAM)—lobectomy • Sacrococcygeal teratoma (SCT)—resection • Meningomyelocele (MMC)—repair • Cervical teratoma—resection
Fetoscopic surgery	• Balloon occlusion of trachea (for congenital diaphragmatic hernia) • Laser ablation of vessels (for twin-to-twin transfusion syndrome) • Cord ligation/division • Cystoscopic ablation of valves (urinary obstruction) • Amniotic bands release
Fetal image-guided surgery	• Amnioreduction/infusion • Fetal blood sampling/intrauterine transfusion • Vascular occlusion procedures (bipolar electrocautery, radiofrequency ablation, interstitial laser ablation, or thrombogenic coil injection) for complex monochorionic twin pregnancies, hydropic lung lesions or sacrococcygeal teratomas • Prenatal stem cell transplantation • Vesico/thoracoamniotic shunts • Balloon dilatation aortic stenosis/stent placement
Exit procedures	• Congenital high airway obstruction (CHAOS) • Removal of the congenital diaphragmatic hernia (CDH) tracheal occlusion balloon • Pulmonary sequestration • Congenital pulmonary airway malformation (CPAM)

(BPS) with pleural effusion may cause mediastinal shift and ultimately fetal hydrops which may lead to fetal demise. Perinatal survival rate in hydropic fetuses with FPE or congenital cystic adenomatoid malformation (CCAM) ranges from 5 to 30%. Fetal intervention for these lesions is aimed at permanently reducing the space-occupying effect of the lesion. Prenatal fetal intervention is primarily indicated in cases with massive pleural effusion and fetal hydrops and is associated with an increased survival rate of up to 65%.[1]

Vesicoamniotic Shunt

Congenital lower urinary tract obstruction (LUTO) occurs in approximately 2.2 per 10000 live births. Causes of LUTO are posterior urethral valve (PUV), mid-urethral hypoplasia, anterior urethral valve, urethral stenosis, ureterocele and urethral agenesis and strictures. LUTO can lead to abnormal renal development. Oligohydramnios resulting from renal impairment between 16 and 24 weeks, is associated with a high prevalence of pulmonary hypoplasia, resulting in high perinatal mortality and morbidity. *In utero*

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percutaneous vesicoamniotic shunt (VAS) is the most common antenatal treatment in these cases to relieve the urinary obstruction by providing bladder drainage and restoring amniotic fluid volume. This leads to increase in prenatal survival. However, a large randomized controlled trial (PLUTO) was conducted comparing VAS versus conservative management of LUTO and concluded that although there was observational evidence for improved perinatal survival, the chance of newborn babies surviving with normal renal function was very low and not improved by VAS.[2]

Though no large randomized controlled trial has been since then, some recent studies report favorable outcome with VAS.[3]

Difference in preprocedure renal function can be a reason for this discrepancy with better outcome in fetuses with functioning kidneys. Assessing fetal renal function, however remains a challenge. Fetal urinary biochemistry, fetal serum beta 2-microglobulin, amniotic fluid amount (oligohydramnios and anhydramnios) and the appearance (hyperechogenic and cystic) of the renal cortex may show promising accuracy for poor postnatal renal function.

Ventriculoamniotic Shunt

Ventriculoamniotic shunts were developed to provide consistent ventricular decompression in cases of hydrocephalus. Though there were few case reports of successful outcome, overall results proved to be poor, often related to undetected central nervous system and noncentral nervous system anomalies, and the shunts failed to provide consistent ventricular decompression because of obstruction or migration.

Recent advances in prenatal diagnosis may allow for the identification of a patient population amenable to *in utero* intervention, namely fetuses with isolated Aqueductal stenosis (AS) diagnosed in the mid-trimester.[4] Also, newer shunt technologies are being developed: ultrasound-guided percutaneous insertion, an anchoring mechanism to prevent migration, a one-way valve to prevent amniotic fluid reflux, and sufficient compliance to avoid cerebral injury.

FETAL LASER THERAPY

Fetal laser therapy utilizes various lasers (most commonly YAG) for vascular coagulation and rarely tissue ablation for conditions described as follows:

Twin-to-twin Transfusion Syndrome

Twin-to-twin transfusion syndrome (TTTS) is a complication of monochorionic twin pregnancy resulting from unbalanced vascular anastomosis between the two fetuses leading to various changes ranging from liquor discrepancy to demise of twins. Laser therapy in the form of ablation of

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the vascular anastomosis between the two fetuses has become the standard treatment for TTTS as it corrects the underlying pathophysiology directly and is seen to have perinatal survival of at least one twin in 81–88% of pregnancies and survival of both twins in 52–54% of pregnancies.[5] Introduction of the 30° fetoscope for anterior placentae has led to significant improvement in survival of both twins.

Laser therapy of TTTS also leads to an improvement in long-term neurodevelopmental outcome. However, laser therapy may lead to complications such as iatrogenic preterm prelabor rupture of membranes, twin anemia–polycythemia sequence (TAPS), reversal or recurrence of TTTS and intrauterine fetal death.

Two techniques are used for this. Either selective coagulation of vascular anastomosis are done or complete equatorial laser ablation is done (Solomon technique). The latter is seen to be associated with a reduction in twin anemia polycythemia sequence (3% vs. 16%) and recurrence of twin-to-twin transfusion syndrome (1% vs. 7%). Perinatal mortality and severe neonatal morbidity is not seen to differ significantly.[6]

Twin Reversed Arterial Perfusion (TRAP)

It can be done by fetoscopic cord ligation of acardiac twin or interstitial laser ablation of acardiac twin vessels. The latter is less invasive than fetoscopic approach.

Fetal Reduction Monochorionic Twin

Since injection of KCL for fetal reduction in monochorionic twin is associated with risk of co-twin demise in view of shared placenta, fetal reduction by cord occlusion can be done by laser therapy. Laser is also associated with lesser risk of co-twin demise than radiofrequency ablation (RFA).

In monoamniotic pregnancies, however, the surviving co-twin still remains at serious risk for intrauterine death (IUD) due to progressive cord entanglement. Cord transection has therefore been recommended to protect the survivor.[7]

Severe IUGR in one Twin of Monochorionic Pair

Single twin death in monochorionic pair can lead to severe neurological sequelae in the surviving twin. In case of severe intrauterine growth restriction (IUGR) in one twin where there is high probability of single twin demise, laser of anastomosing vessels can be done. It may not benefit the growth restricted fetus, however, it protects the normal fetus from neurological damage even when the growth restricted twin dies.

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Fetal Thoracic Mass Lesions

Bronchopulmonary sequestration (BPS) is a rare malformation of the respiratory tract consisting of a mass of bronchopulmonary tissue that is separate from the tracheobronchial tree and fed by a separate systemic artery.

Ultrasound-guided laser coagulation of the feeding artery is done leading to regression of the mass and clinical improvement.

In cases with massive pleural effusion, 'vascular' laser ablation of the feeding vessel seems to be more effective than pleuroamniotic shunting, with fewer complications.

However, outcome of laser ablation in treating CPAM have so far been disappointing, with survival rates below 50% hence distinction between CPAM and BPS is of utmost importance when prenatal intervention is considered.[8]

Lower Urinary Tract Obstruction

The ideal management of fetuses with severe LUTO continues to be one of the most challenging subjects in the field of fetal intervention. Although experience with percutaneous fetal cystoscopy and endoscopic release of PUV has been limited, this alternative may have advantages over other prenatal fetal therapies. Fetoscopy has a role to play in the diagnostic evaluation of fetal obstructive uropathies. Treatment of PUV with direct fulguration of the valves, leads to a more physiological treatment of the obstruction which may perhaps prevent postnatal renal dysfunction. There is still a significant risk of progression to renal failure pre- or postnatally. Currently no randomized trial is available.

Chorioangioma

There are limited interventions available for the management of large, symptomatic placental chorioangiomas. They can be treated with endoscopic laser coagulation of the feeding vessels, however fetal bleeding, exsanguination, and death are potential complications of the procedure; therefore, this technique should be used with caution.

Sacrococcygeal Tumor

Percutaneous ultrasound-guided intratumor laser ablation has been used for treatment of sacrococcygeal tumor. The reported survival from intratumor laser or radiofrequency ablation is similar (about 50%).[9]

Vasa Previa Type 2

Vasa previa is a rare placental disorder in which fetal blood vessels, unsupported by underlying placenta, cross-over the cervical internal os; type 1

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vasa previa is attributed to a velamentous cord insertion near the cervix while type 2 vasa previa occurs when connecting vessels of a bi or multilobed placenta cross-over internal cervical. Laser coagulation of vessels is used in type 2 in the presence of premature contractions or progressive cervical shortening with risk of bleeding and fetal exsanguination. Initial experience suggests that fetoscopic laser ablation of type 2 vasa previa appear to be safe and feasible.[10]

Amniotic Band Syndrome

Amniotic bands may cause entanglement of fetal parts or umbilical cord and lead to multiple fetal defects ranging from constriction of limb to fetal death. Lysis of the amniotic sheets is performed using laser dissection which helps in prevention of further damage and possibly reversal of edema. Umbilical cord constriction and severe limb constriction with an abnormal distal Doppler or an absent vascular flow are indications for performing a fetoscopy and laser.[11]

Fetal Cardiac Lesions

A novel use of laser is seen in cardiac lesions. A case of hypoplastic left heart syndrome (HLHS) with intact atrial septum (50% neonatal mortality from profound hypoxemia since oxygenated blood cannot exit the left atrium) was reported in which thulium laser was used to vaporize a hole through the thick septum from a left atrial approach (29w5d of gestation). The needle used to deliver the stent was then easily advanced over the fiber into the right atrium. This was followed by an uncomplicated spontaneous vaginal delivery, a baby girl was delivered at 40w1d. She underwent Norwood procedure postdelivery.[12]

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