

Intensive Care Nursery House Staff Manual

Fetal Therapy

DEFINITION: A therapeutic intervention for the purpose of correcting or treating a fetal anomaly or condition. In almost every case, the fetus is at risk of intrauterine death from the abnormality.

INTRODUCTION: UCSF has utilized or pioneered several types of fetal therapy. These interventions are limited to a few specific conditions, where therapy has either proven beneficial or is under investigation. Largely as a result of the Fetal Treatment Program, the perinatal patient population at UCSF (maternal and neonatal) is unique with regard to the number of fetuses and newborns with unusual or rare conditions. These patients are discussed at the weekly multidisciplinary Fetal Treatment Meeting (Tuesday, 1:00 PM).

PATIENT SELECTION: For all interventions, mothers are counseled extensively by appropriate specialists (*e.g.*, Pediatric Surgeons, Perinatologists, Neonatologists, Anesthesiologists, Ultrasonographers, Neurosurgeons, Social Workers) with regard to the nature of the condition, possible risks and benefits, alternative treatments, and potential outcomes. The most common conditions for which fetal interventions are considered are:

Erythroblastosis Fetalis: In very severe cases, fetal intrauterine transfusion is performed to treat the hemolytic anemia. For further information, see the section on Hemolytic Disease of the Newborn (P. 121).

Congenital Diaphragmatic Hernia (CDH): The major causes of morbidity and mortality with CDH are pulmonary hypoplasia and persistent pulmonary hypertension. In experimental animals, fetal tracheal occlusion stimulates lung growth by lung distension with fetal lung fluid. Although fetal tracheal occlusion is no longer used for most cases of CDH, it is occasionally considered for the most severe cases of CDH for whom survival is <10%. Fetuses with tracheal occlusion must be delivered by EXIT procedure (partial delivery of the fetus, removal of the tracheal occlusion, administration of surfactant and institution of assisted ventilation while the infant is still on placental support).

Urinary Tract Obstruction: Complete obstruction of the fetal urinary tract results in severe renal damage as well as pulmonary hypoplasia from severe oligohydramnios. Despite early enthusiasm for fetal decompression of the urinary tract, fetal intervention has seldom been beneficial and is now rarely performed.

Fetal Tumors Causing Hydrops Fetalis: When the relatively rare fetal tumors, congenital cystic adenomatoid malformation of the lung (CCAM) and sacrococcygeal tumor (SCT), are associated with hydrops fetalis, fetal mortality approaches 100%. These tumors cause hydrops by either venous obstruction due to

mediastinal shift (CCAM) or high output heart failure (SCT). Operative removal of these tumors has resulted in survival of ~50% of the affected fetuses.

Twin-Twin Transfusion Syndrome (TTTS): Monochorionic twins have a high frequency of placental vascular shunts that may lead to one twin (donor) over-perfusing the other (recipient). Complications include oligohydramnios and growth retardation (donor), polyhydramnios and hydrops fetalis (recipient), and fetal death. Currently, UCSF is participating in an NIH sponsored multi-center controlled trial of fetal intervention (amnio-reduction, obliteration of shunt vessels).

Meningomyelocele (MMC): Currently, UCSF is participating (with Children's Hospital of Philadelphia and Vanderbilt University) in an NIH-sponsored multi-center controlled trial of fetal repair of MMC. This trial is notable because fetal repair of MMC is the first fetal intervention for a condition that is not life-threatening to the fetus. The primary outcome variable is the need for ventriculo-peritoneal shunt as treatment for hydrocephalus, which occurs in approximately 80% of infants after post-natal repair of MMC.

Other Conditions: It is likely that other conditions will become subjects of attempted fetal correction or treatment. Currently, consideration is being given to fetal intervention for certain cases of hypoplastic left heart syndrome.

NEONATAL CARE: A still unsolved complication of most fetal interventions is premature birth. The degree of prematurity varies with the condition, the type of fetal intervention, and the gestation at which it was performed. In some cases (*e.g.*, TTTS), no specific neonatal care is needed other than care of the premature infant. In other cases (*e.g.*, CDH), the infant will require intensive and complex resuscitation necessitating a large neonatal team.