

Contents

Foreword: Fetal and Neonatal Surgery: Then and Now **xv**

Lucky Jain

Preface: Advances and Updates in Fetal and Neonatal Surgery **xix**

KuoJen Tsao and Hanmin Lee

The Rearing of Maternal–Fetal Surgery: The Maturation of a Field from Conception to Adulthood **799**

Caressa Chen, Lauren L. Evans, and Michael R. Harrison

Maternal–fetal surgery is fraught with inherent controversy from within the medical community and general public. Despite these challenges, the field of maternal–fetal surgery evolved into an international enterprise. Carefully nurtured by pioneers with foresight and resilience, the field navigated ethical dilemmas with rigorous scientific methodology, collaboration, transparency, and accordance. These central pillars are consistent throughout the brief but momentous history of maternal–fetal surgery, serving as the catalyst for its success. The maturation of fetal intervention is an exemplar of technological innovation propelling clinical innovation, as well as a celebration of mastering the delicate balance between caution and optimism.

Molecular and Cellular In Utero Therapy **811**

Cara L. Berkowitz, Valerie L. Luks, Marcelina Puc, and William H. Peranteau

Significant advances in maternal–fetal medicine and gene sequencing technology have fostered a new frontier of in utero molecular and cellular therapeutics, including gene editing, enzyme replacement therapy, and stem cell transplantation to treat single-gene disorders with limited post-natal treatment strategies. In utero therapies take advantage of unique developmental properties of the fetus to allow for the correction of monogenic disorders before irreversible disease pathology develops. While early preclinical studies in animal models are encouraging, more studies are needed to further evaluate their safety and efficacy prior to widespread clinical use.

Fetal and Neonatal Anesthesia **821**

Marla B. Ferschl and Ranu R. Jain

Anesthesia for fetal and neonatal surgery requires subspecialized knowledge and expertise. Attention to important anatomic, physiologic, and metabolic differences seen in pregnancy and at birth are essential for the optimal care of these patients. Thorough preoperative evaluations tailored intraoperative strategies and careful postoperative management are critical when devising the anesthetic approach for each of these cases.

Fetal Repair of Neural Tube Defects

835

Su Yeon Lee, Ramesha Papanna, Diana Farmer, and KuoJen Tsao

Myelomeningocele is the most common congenital neurologic defect, and the only nonlethal disease addressed by fetal surgery. A randomized control trial has established amelioration of the Arnold–Chiari II malformation, reduced ventriculoperitoneal shunt rate, and improvement in distal neurologic function in patients that receive in utero repair. Long-term follow-up of these school-age children demonstrates the persistence of these effects. The use of stem cells in fetal repair is being investigated to further improve distal motor function.

Fetal Therapy for Renal Anhydramnios

849

Jena L. Miller, Ahmet A. Baschat, and Meredith A. Atkinson

The most severe forms of congenital anomalies of the kidney and urinary tract present in fetal life with early pregnancy renal anhydramnios and are considered lethal due to pulmonary hypoplasia without fetal therapy. Due to the high rate of additional structural anomalies, genetic abnormalities, and associated syndromes, detailed anatomic survey and genetic testing are imperative when stratifying which pregnancies are appropriate for fetal intervention. Restoring amniotic fluid around the fetus is the principal goal of prenatal treatment. The ongoing multi-center Renal Anhydramnios Fetal Therapy (RAFT) trial is assessing the safety and efficacy of serial amnioinfusions to prevent pulmonary hypoplasia so that the underlying renal disease can be addressed.

In Utero Therapy for Congenital Diaphragmatic Hernia

863

Marisa E. Schwab, Hanmin Lee, and KuoJen Tsao

Congenital diaphragmatic hernia is an anomaly that is often prenatally diagnosed and spans a wide spectrum of disease, with high morbidity and mortality associated with fetuses with severe defects. Congenital diaphragmatic hernia is thus an ideal target for fetal intervention. We review the literature on prenatal diagnosis, describe the history of fetal intervention for congenital diaphragmatic hernia, and discuss fetal endoscopic tracheal occlusion and the Tracheal Occlusion To Accelerate Lung growth trial results. Finally, we present preclinical studies for potential future directions.

Updates in Neonatal Extracorporeal Membrane Oxygenation and the Artificial Placenta

873

Brianna L. Spencer and George B. Mychaliska

Extracorporeal life support, initially performed in neonates, is now commonly used for both pediatric and adult patients requiring pulmonary and/or cardiac support. Data suggests the clinical feasibility of Extracorporeal Membrane Oxygenation for premature infants (29–33 weeks estimated gestational age [EGA]). For extremely premature infants less than 28 weeks EGA, an artificial placenta has been developed to recreate the fetal environment. This approach is investigational but clinical translation is promising. In this article, we discuss the current state and advances in neonatal and “preemie Extracorporeal Membrane Oxygenation” and the development of an artificial placenta and its potential use in extremely premature infants.

Surgical Management of Congenital Diaphragmatic Hernia 893

Matthew T. Harting and Tim Jancelewicz

Congenital diaphragmatic hernia (CDH) is a challenging surgical disease that requires complex preoperative, perioperative, and postoperative care. Survival depends on successful reduction and repair of the defect, and numerous complex decisions must be made regarding timing and preparation for surgery. This review describes the challenges and controversies inherent to surgical CDH care and provides recommendations for management based on the most recent evidence.

Management of Congenital Lung Malformations 907

Brittany N. Hegde, KuoJen Tsao, and Shinjiro Hirose

Congenital lung malformations represent a spectrum of lesions, each with a distinct cause and tailored clinical approach. This article will focus on the following malformations: congenital pulmonary airway malformations, formally known as congenital cystic adenomatoid malformations, bronchopulmonary sequestration, congenital lobar emphysema, and bronchogenic cyst. Each of these malformations will be defined and examined from an embryologic, pathophysiologic, and clinical management perspective unique to that specific lesion. A review of current recommendations in both medical and surgical management of these lesions will be discussed as well as widely accepted treatment algorithms.

Advances in Complex Congenital Tracheoesophageal Anomalies 927

Somala Mohammed and Thomas E. Hamilton

Esophageal atresia with or without tracheoesophageal fistula and tracheobronchomalacia encompass 2 of the most common complex congenital intrathoracic anomalies. Tailoring interventions to address the constellation of problems present in each patient is essential. Due to advances in neonatology, anesthesia, pulmonary, gastroenterology, nutrition and surgery care for patients with complex congenital tracheoesophageal disorders has improved dramatically. Treatment strategies tailored to the individual patient needs are best implemented under the aegis of a comprehensive longitudinal multidisciplinary care team.

Abdominal Wall Defects: A Review of Current Practice Guidelines 943

Alyssa R. Mowrer, Daniel A. DeUgarte, and Amy J. Wagner

The 2 most common congenital abdominal wall defects are gastroschisis and omphalocele. Gastroschisis is a defect in the abdominal wall with exposed abdominal contents. Mortality rates are low but lengths of stay are often prolonged by bowel dysmotility and other intestinal abnormalities in complicated cases. Omphalocele is a defect through the umbilical cord with herniated abdominal contents covered by a sac. It is associated with other genetic abnormalities and other anomalies that can lead to significant morbidity and mortality. Prenatal diagnosis in both conditions allows for improved prenatal consultation and coordinated perinatal care to improve clinical outcomes.

Review of Necrotizing Enterocolitis and Spontaneous Intestinal Perforation Clinical Presentation, Treatment, and Outcomes

955

Laura A. Rausch, David N. Hanna, Anuradha Patel, and Martin L. Blakely

The Necrotizing Enterocolitis Surgery Trial (NEST) highlights the importance of distinguishing necrotizing enterocolitis (NEC) from spontaneous intestinal perforation (SIP) when developing surgical treatment plans. Further research is needed to increase the accuracy of this distinction, but even with our current abilities to do this initial laparotomy appears to be optimal for infants with presumed NEC. The preferred initial operation for those with SIP is more equivocal. Rates of NEC are likely decreasing slowly, whereas those with SIP are not. New imaging modalities, especially ultrasound, are becoming more useful but require more detailed investigation. Understanding the mechanisms causing these two conditions remains of paramount importance.

Advances in the Management of the Neonate Born with an Anorectal Malformation

965

Sebastian K. King and Marc A. Levitt

Anorectal malformations occur in 1 in 3000 to 5000 children, and present with a marked variety in type and severity. Most of the malformations are diagnosed in the early neonatal period, as an antenatal diagnosis remains relatively elusive. Following diagnosis, an accurate assessment and focused management is crucial to reduce the potential for morbidity and mortality. This review focuses on the investigation and management of newborns with anorectal malformations, and the introduction of novel assessment tools for the more complex malformation types.

Advances in the Treatment of Neonatal Biliary Disease

981

Sarah Mohamedaly and Amar Nijagal

This article discusses current standard of care in neonatal biliary disease, particularly management of biliary atresia and choledochal cysts. It highlights surgical considerations, guidelines for adjuvant therapies, and promising therapeutic options that are under investigation.