Welcome!

May 24, 2022

The webinar will begin at 6 p.m. CT

Agenda for the evening

- 6:00 p.m. | **Welcome** by Erika Peterson, MD
- 6:05 p.m. | ***New Management Algorithm for Fetal and Neonatal Bladder Exstrophy*** by John Kryger, MD
- 6:35 p.m. | ***Equipoise for and Status of the Gastroschisis Outcomes of Delivery (GOOD) Study*** by Amy Wagner, MD
- **7:05 p.m. | *Prenatal Predictors of Outcomes in Congenital Heart Disease*** by David Saudek, MD
- **7:45 p.m. | Wrap-up** by Erika Peterson, MD
- **Q&A sessions to follow after each presentation.**

Take a tour

- https://www.youtube.com/watch?v=dmqEKSACGKA

Physician Liaisons are here for you

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Margie Berg
Sr. Physician Liaison
mberg2@chw.org

Southeast Wisconsin

- **Serve as a link between Children's Wisconsin and referring providers**
- **Provide information about Children's Wisconsin services and programs, including continuing education opportunities**
- **Facilitate solutions for referral and communication issues**

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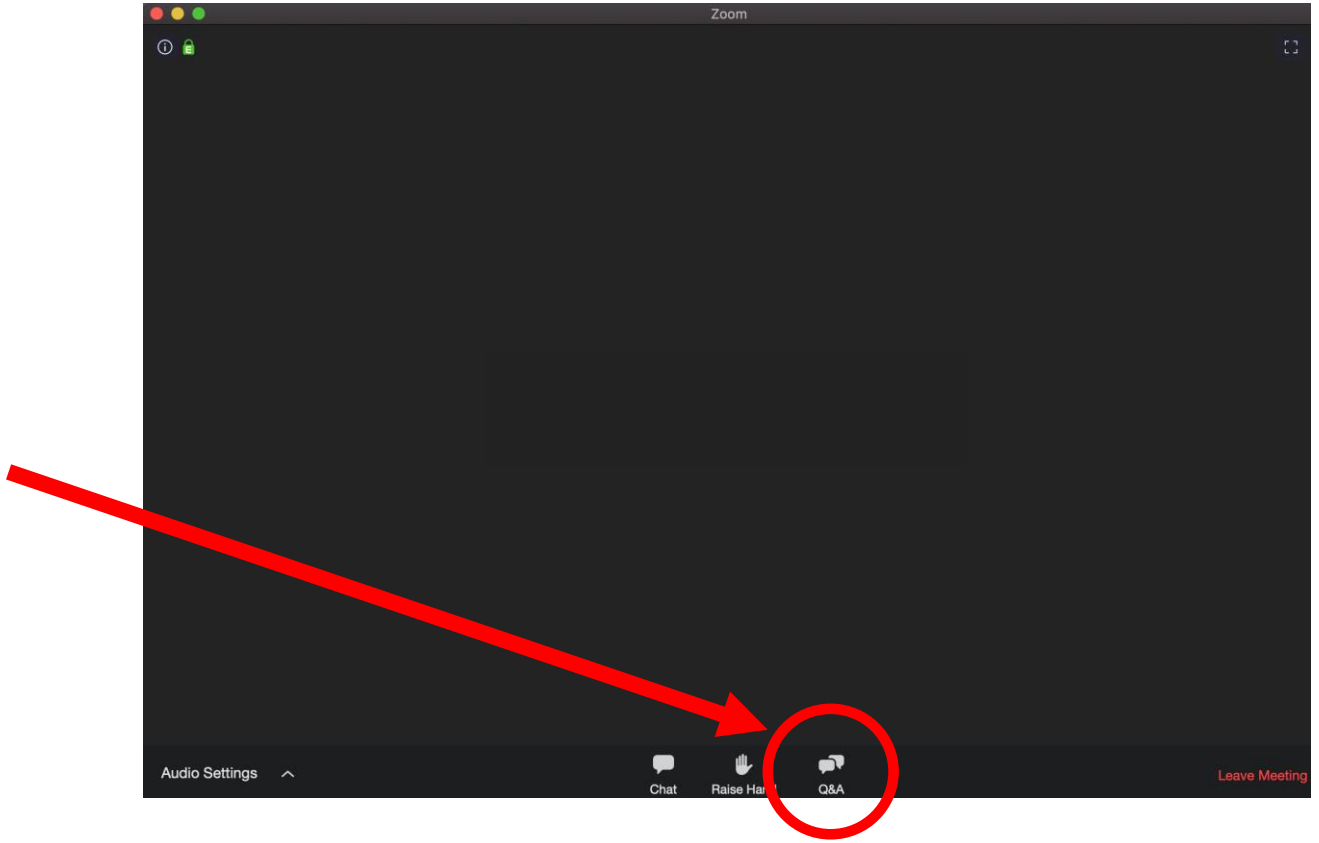
Asking the presenter a question

At any time during the lecture, you can submit your question by **clicking the Q&A icon** at the bottom of your screen



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John Kryger, MD

- Medical director of urology at Children's Wisconsin since 2009
- Chief and professor of pediatric urology at the Medical College of Wisconsin
- Board certified in pediatric urology
- Earned his medical degree from University of Wisconsin Medical School - Madison
- Completed his pediatric urology fellowship at Children's Hospital of Michigan
- Dr. Kryger sees patients at the Milwaukee Campus, Appleton Clinic, and Delafield Clinic

Advancements in Fetal and Neonatal Bladder Exstrophy management

John V. Kryger
Professor and Chief of Pediatric Urology
Children's Hospital of Wisconsin
Milwaukee, WI

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Disclosures

- In accordance with ACGME policy:
  I have no financial disclosures

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Outline

- Define and describe Bladder exstrophy
- Explain the need for improved care
- Describe a new philosophy to care in Multi-disciplinary Bladder Exstrophy Program
  - One of 12 Centers of Excellence in US
- Describe how newborn care of bladder exstrophy has changed in the nursery.

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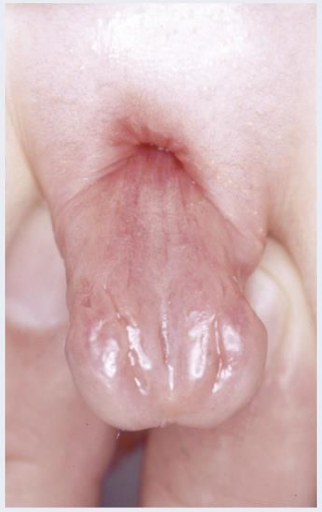
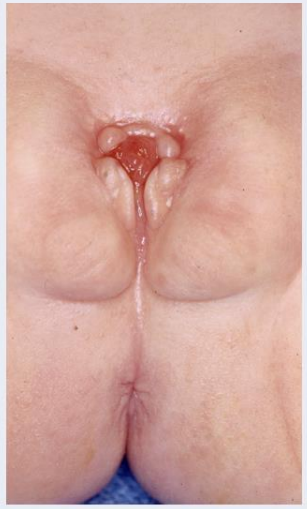
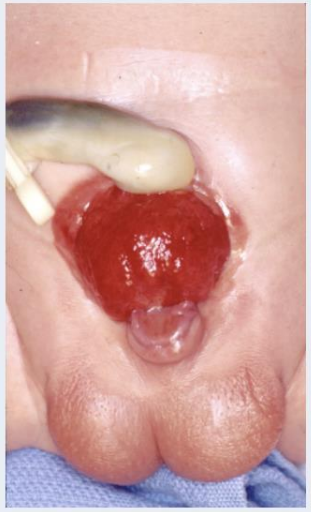
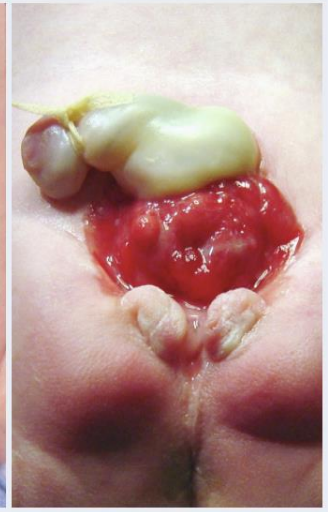
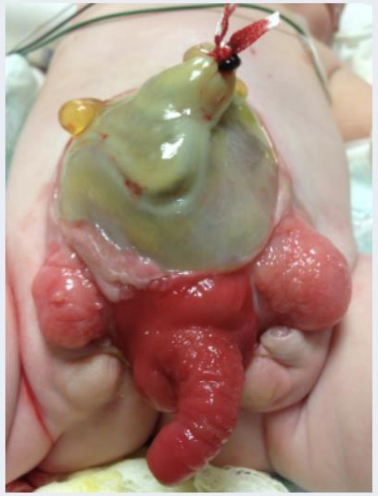
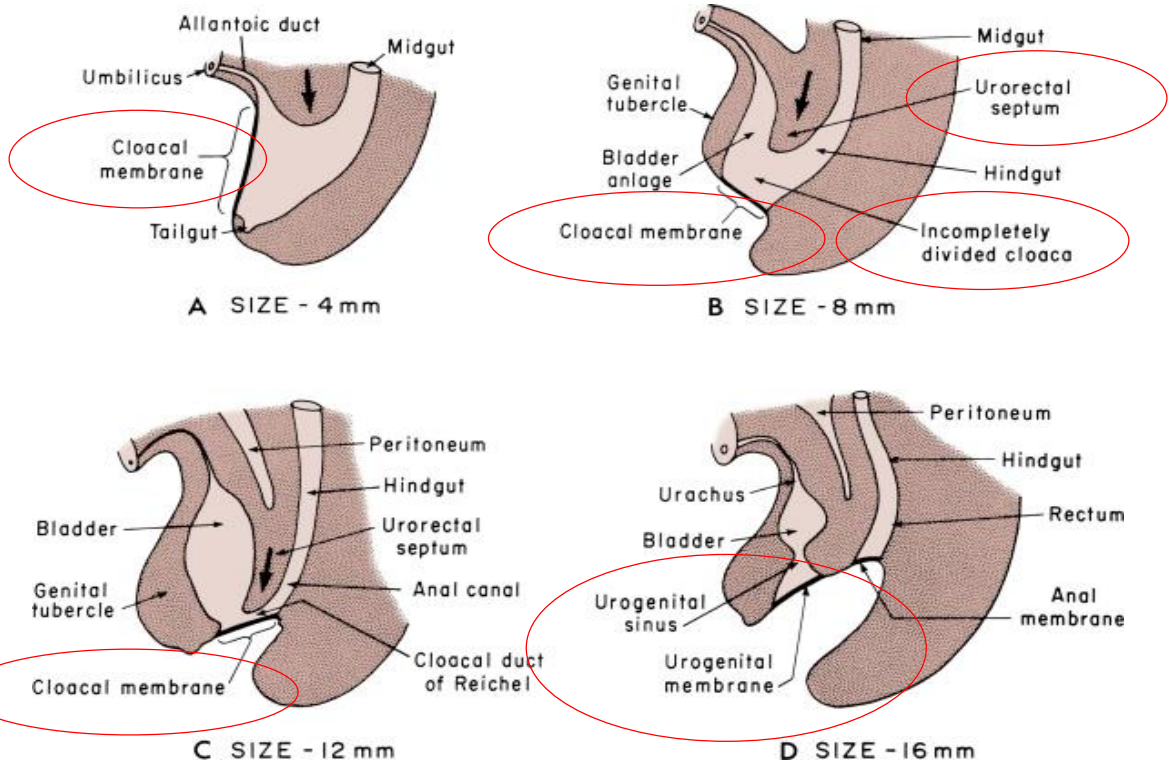
Bladder Exstrophy

- Bladder exstrophy is a spectrum
- Wisconsin birth rate is 60,000 per year (2020)

Epispadias	Bladder Exstrophy	Cloacal Exstrophy
1:150,000	1:40,000	1:400,000
Male  Female 	Male  Female 	

Premature rupture of cloacal membrane

- *Mesenchymal ingrowth* is critical to stability of the membrane
  - As the urorectal septum separates the GU and GI tracts
  - As pelvic organs, abdominal wall, pelvic bones mature
  - As external genitalia develop
- Premature membrane rupture results in **HERNIATION** of pelvic organs



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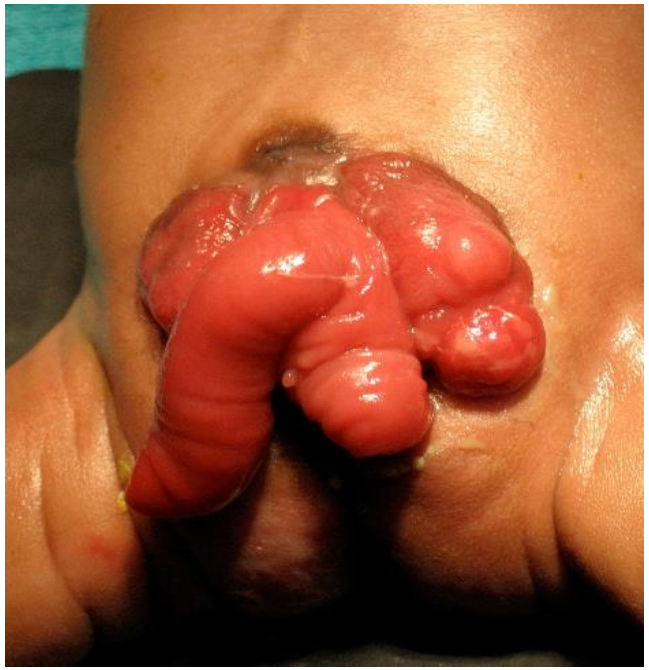
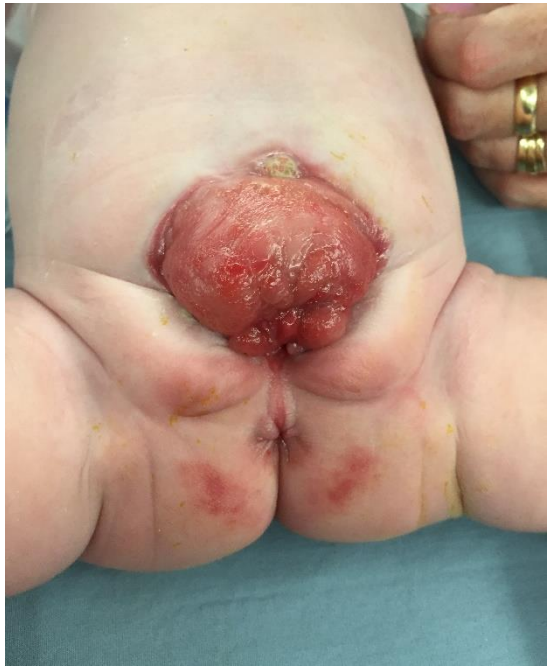
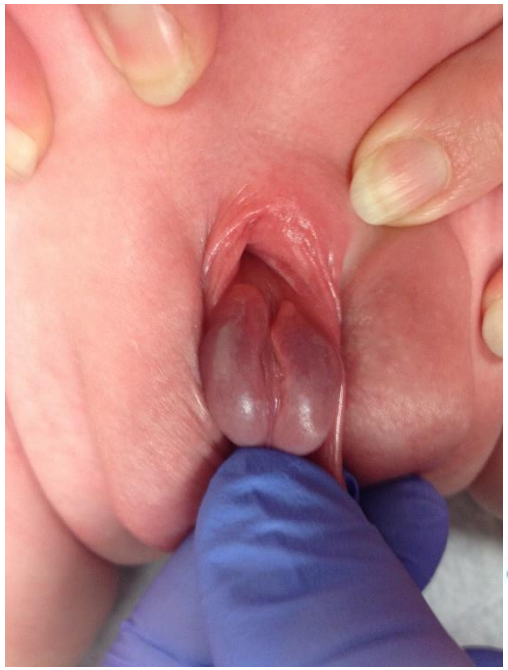
Exstrophy Etiology

Timing has major impact on the severity

- 4-6 weeks Cloacal exstrophy
- 6-12 weeks Classic bladder exstrophy
- 12-14 weeks Epispadias







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Bladder Exstrophy | Wide spectrum of Anomalies

Male Genital Defects

- Shortened penis
- Congenital dorsal chordee
- Neurovascular displacement

Female Genital Defects

- Anterior displaced/ short vagina
- Bifid clitoris

Urinary Tracts Defects

- Vesico-ureteral reflux
- Variety of renal anomalies
- Issues of bladder development are certainly complex

Musculoskeletal Defects

- Separation of the pubic symphysis
- Outward rotation/ shortening pubic rami

Abdominal Wall Defects

- Inferior abdominal wall muscle defect
- High incidence of inguinal hernia

Neurologic Defects

- Spinal cord anomalies

Anorectal Defects

- Anterior displacement of anus
- Pelvic floor muscle deficiency

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Prenatal Diagnosis

- Accurate prenatal diagnosis has major impact on successful counseling and setting expectations
    - Guides postnatal care and avoids urgent transfers of infant and separation from mother
- Only 10-32% of exstrophy babies have prenatal diagnosis (1)

1. Goyal A, Fishwick J, et al. Antenatal diagnosis of Bladder/cloacal exstrophy: Challenges and possible solution. J Ped Urol 2012

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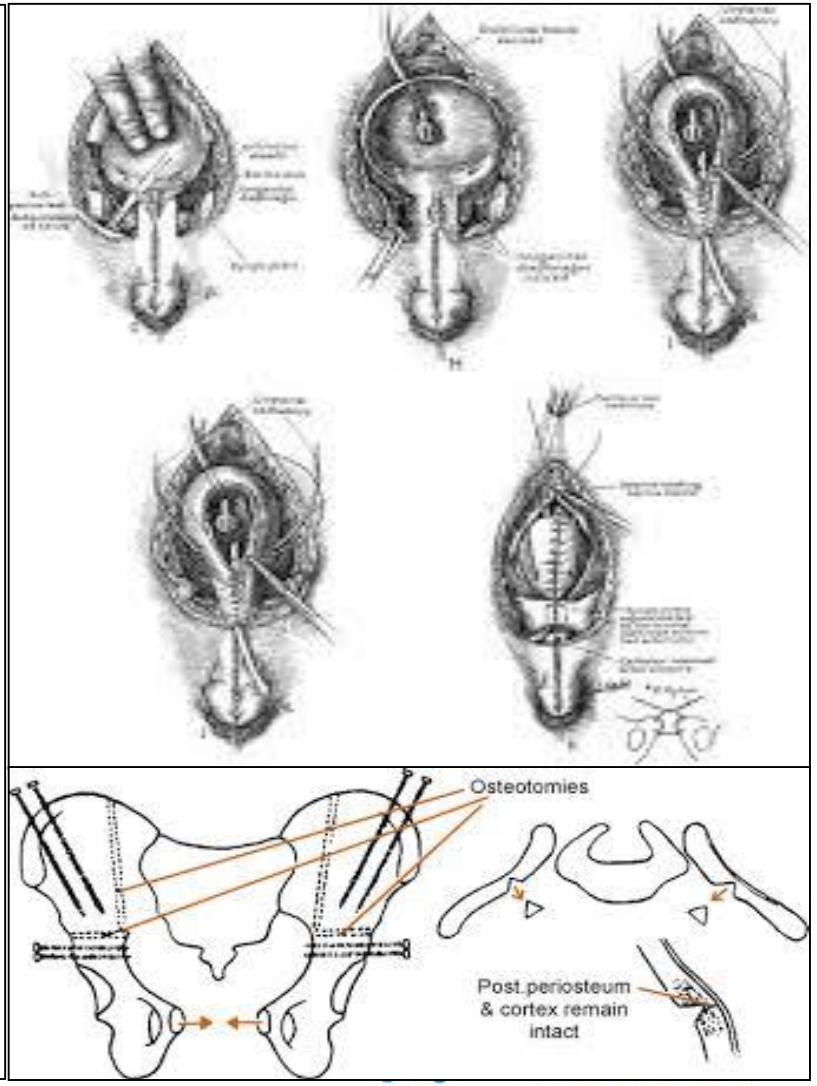
Fetal Imaging

- Fetal US findings:
  - Absence of bladder
  - Widened pubic diatasis
  - Low lying umbilicus
  - Presence of omphalocele is key differention: BE vs CE
    - Can be difficult to distinguish true omphalocele from bulging bladder plate
    - Sensitivity distinguishing BE from CE (1)
      - fUS 69% fMRI 83%

1. Weiss D, Kryger J, et al. Key anatomic findings on fetal ultrasound and MRI in the prenatal diagnosis of bladder and cloacal exstrophy. J Ped Urol 2020

Two Approaches to surgery
- Slow progress over 75 years

1. **Staged Repair**
   - Primary Closure of the bladder and abdomen at birth
   - Epispadias repair at 18 months.
   - Bladder Neck reconstruction at 5-8 years of age
2. **Complete Primary Repair as infant (one stage CPRE)**
   - Closure of exstrophy
   - repair of epispadias
   - bladder neck reconstruction
   - osteotomies



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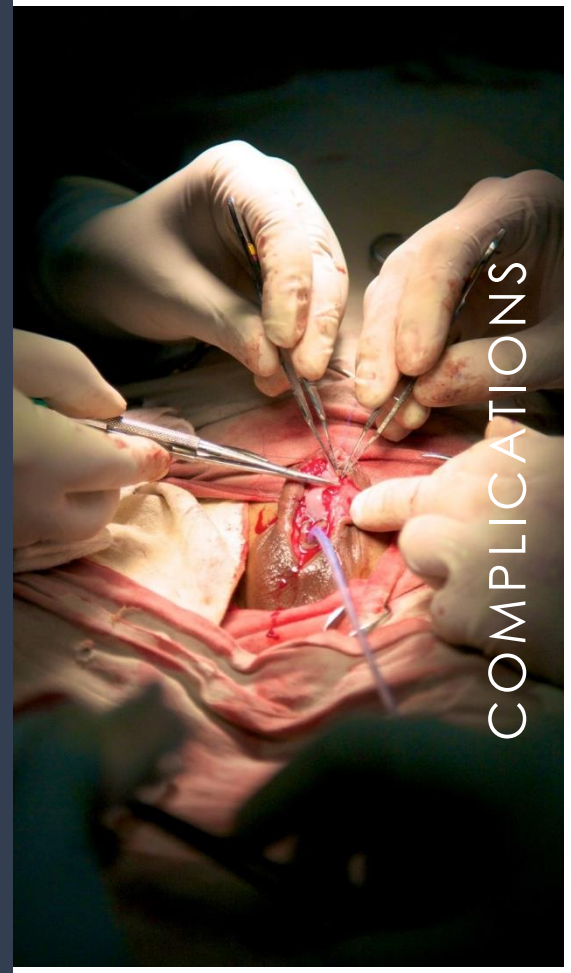
Pitfalls of this Surgery

- ***Are common and sometimes devastating***

DEHISCENCE & FISTULA	PENILE INJURY, EVEN LOSS	UPPER URINARY TRACT DAMAGE
URINARY TRACT INFECTIONS	LONG-TERM URINARY INCONTINENCE	BLADDER AUGMENTATION



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The Complete Primary Repair of Exstrophy (CPRE)

- **Complete Primary Repair was innovation advanced by Dr Mitchell**
- Complete disassembly of penile corpora and urethra and glans
- More aggressive Bladder neck dissection and reconstruction

- Continence 80% among children > 4 yo
- Long-term Continence 29/39 (74%) among children > 4 yo

Critical to his philosophy

1. With proper dissection and reconstruction, the anatomy of the herniated structures can be restored better than in the past
2. Early cycling of the bladder is critical to bladder development

1. Grady, Mitchell J.Urol:162(4), 1415-1420 **October 1999**,
2. Schnooravorian, Grady, Joyner, Mitchell J Urol 180:1615-1620. **October 2008**

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THE NEW YORKER

FICTION SCIENCE & TECH BUSINESS HUMOR MAGAZINE VIDEO

Personal Best

Top athletes and singers have coaches. Should you?

BY ATUL GAWANDE

- Coaches for:
  - Athletes
  - Musicians
- Why not surgeons?

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Coaching

- **Bladder Exstrophy** is an excellent model for surgical coaching.
  - Given **rarity** of this disease gaining necessary **surgical experience** is challenging.
  - The complexity of disease requires **mastery of surgery**

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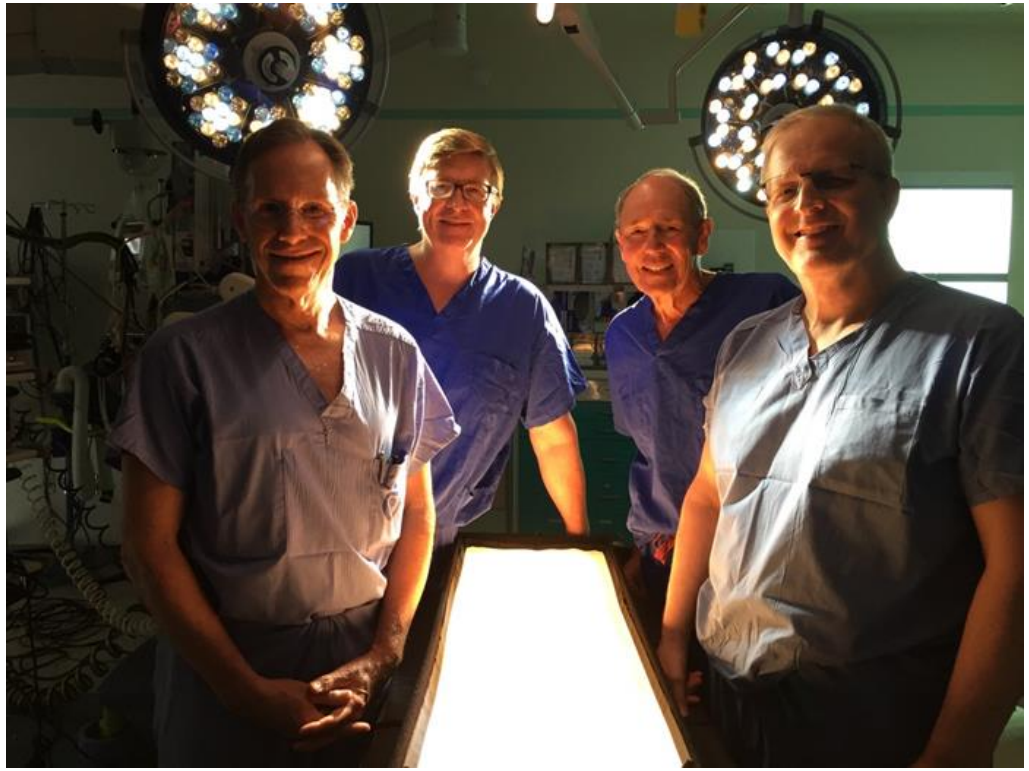
Multi-institutional Bladder Exstrophy Consortium (MIBEC)

- Founded in 2012 to enhance the experience with this rare and complex disease
  - Doug Canning,
    Children's Hosp Philadelphia
  - Joe Borer,
    Boston Children's
  - John Kryger,
    Childrens Wisconsin
  - Mike Mitchell,
    Coach and Mentor

MIBEC
Multi Institutional Bladder Exstrophy Consortium



Why form a consortium?

- Refine and standardize the Mitchell technique of CPRE
- **Reproduce accomplishments reported by Mitchell**
- **Develop an accurate high-volume database**
  - short- and long-term clinical outcomes
  - surgical outcomes
- Develop training and educational resources for Pediatric urologists

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Collaboration

"The collaboration is a mechanism to learn from each other's expertise toward promoting the main goal of optimizing care and outcome for rare and complex patients with exstrophy spectrum."

Mike Mitchell, MD

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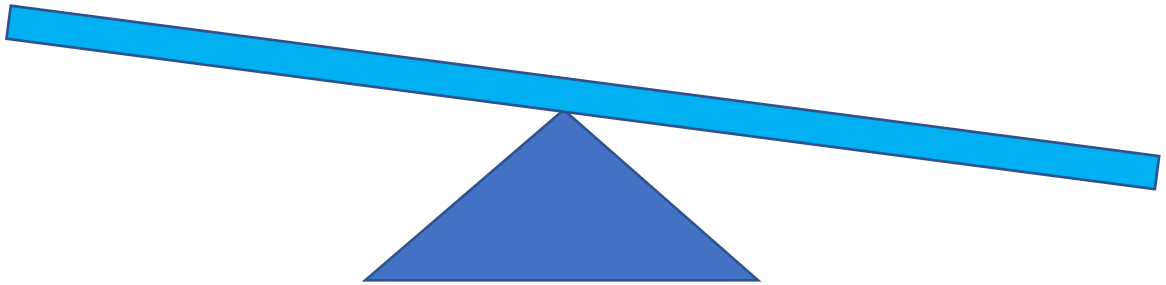
Advancements of MIBEC approach

Immediate Closure 1 day

- Pelvic bone flexibility
- Less bladder irritation
- Early completion for family

Elective Closure 2-3 mos

- Assemble surgery team
- Mini-puberty robust genital tissue
- Better family bonding
- Time for Nutrition and nursing
- Family counselling and preparation



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MIBEC Collaboration: 8 surgeons

MIBEC February 2013 – May 2022

Primary CPRE in **97** patients:

- Male: 61
- Female: 36

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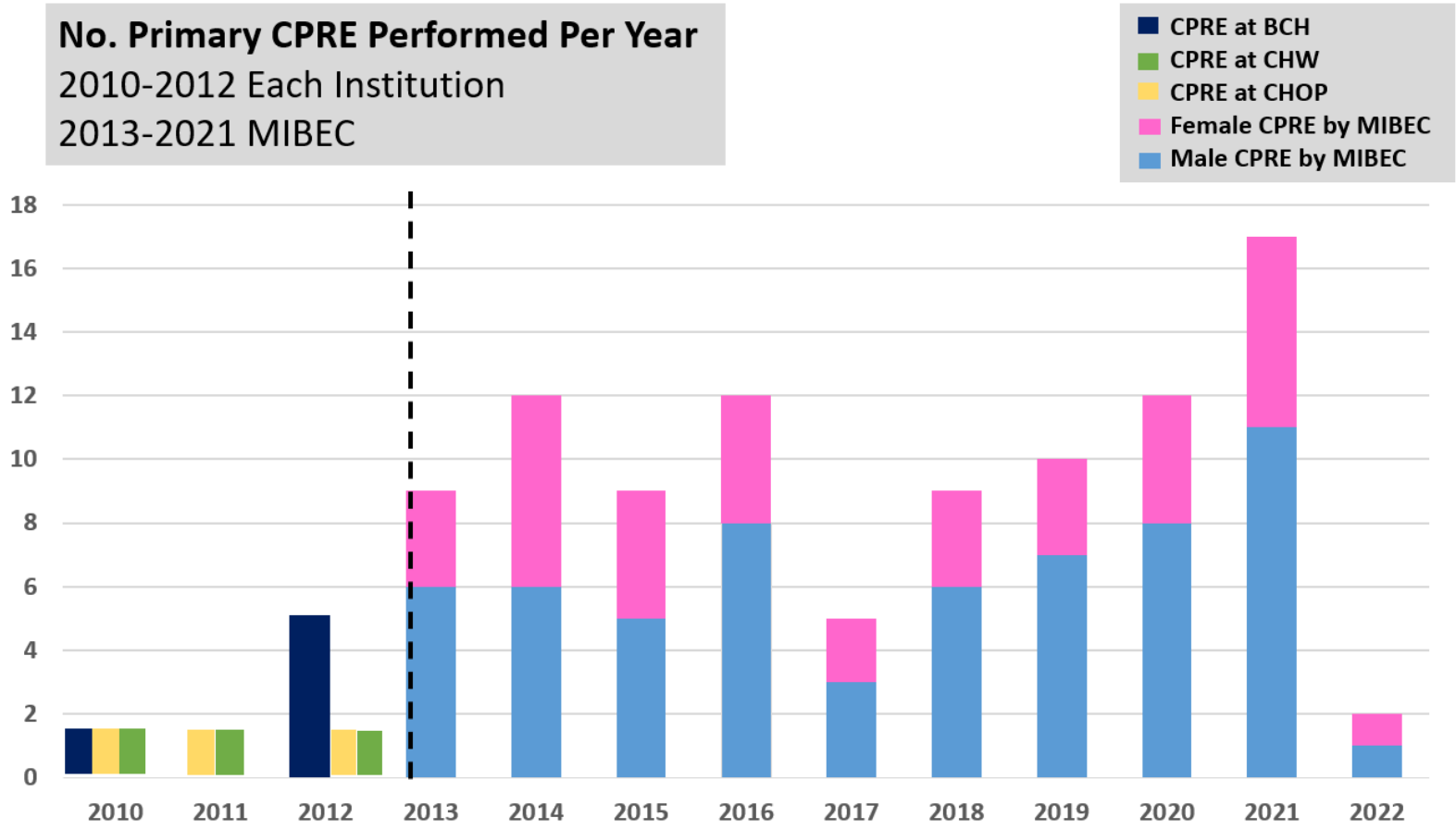
Multi Institutional Bladder Exstrophy Consortium

No. Primary CPRE Performed Per Year
2010-2012 Each Institution
2013-2021 MIBEC



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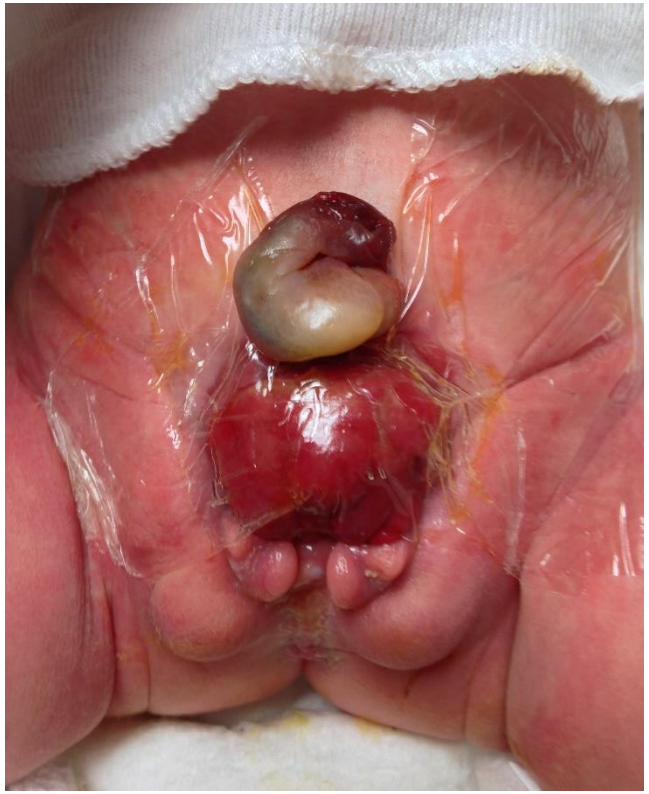
Newborn Care is important esp. when closure delayed

- Bladder mucosa is very sensitive
  - Tie umbilical cord with silk to avoid clamp irritation to bladder mucosa
  - Cover mucosa with **tegaderm** to protect from diaper
- Parent counseling and education
  - Critical to parents on many levels.
  - Takes time to do it well… time well spent



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Newborn Care of Classic Bladder Exstrophy

- Not as involved as most people expect
  - Children are generally robust and healthy at birth
- **Pediatric Urology consult is important**
  - No need to transfer patient
  - No separation of mother/ infant
- No special needs except covering the exposed bladder with tegaderm or press n' seal
- Discharge to home directly from nursery is expected in the usual time frame.
- Appt with Pediatric Urology Center of Excellence within 1 week, preferably in person, but can be telemedicine if great distance

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Newborn Care of Classic Bladder Exstrophy

- Imaging at the Urology visit (can be done prior to discharge from nursery as option)
  - Renal US
  - Pelvic x-ray
- No labs
- No antibiotic prophylaxis
- No restrictions on diapering or bathing or activities
- Essentially normal newborn care

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Patient Satisfaction

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Publications

JAMA Surgery | Original Investigation

The International Bladder Exstrophy Consortium
A Model for Sustained Collaboration to Address the Unmet Global Burden of Surgical Disease

Rakesh S. Joshi, MD; Dhirendra Shrivastava, MD; Richard Grady, MD; Anjana Kundu, MD; Jaishri Ramji, MD; Pramod P. Reddy, MD; Joao Luiz Pippi-Salle, MD; Jennifer R. Frazier, MPH; Douglas A. Canning, MD; Aseem R. Shukla, MD

THE JOURNAL of UROLOGY®
Official Journal of the American Urological Association

An Initial Report of a Novel Multi-Institutional Bladder Exstrophy Consortium: A Collaboration Focused on Primary Surgery and Subsequent Care

Joseph G. Borer, Evalynn Vasquez, Douglas A. Canning, John V. Kryger, Michael E. Mitchell

Journal of Pediatric Urology
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Article in Press

Short-term outcomes of the multi-institutional bladder exstrophy consortium: Successes and complications in the first two years of collaboration☆

J.G. Borer, E. Vasquez, D.A. Canning, J.V. Kryger, A. Bellows, D. Weiss, T. Groth, A. Shukla, M.P. Kurtz, M.E. Mitchell

DOI: http://dx.doi.org/10.1016/j.jpurol.2017.01.006

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Facilitates a Skilled Multidisciplinary Team

- Pediatric Urology
- Pediatric Orthopedics
- Pediatric Anesthesia
- Pediatric Intensive care
- Pediatric Psychology
- Pediatric Pain specialists
- Pediatric Nurses

MIBEC
Multi Institutional Bladder Exstrophy Consortium

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MIBEC – Early Benefits

- Consistent Team of Surgeons
  - Better teaching
  - Safer surgical outcomes
  - Collective wisdom of senior surgeons
- Evolution of the procedure
  - Improved dissection of urethra
  - Better dissection and reconstruction of the bladder sphincter
  - Improved reconstruction of both male and female genitalia
  - Role of osteotomies and spica cast
- Outcomes
  - Improved continence with voiding in normal fashion
  - Great reduction in number of bladder augmentations
  - National recognition, teaching videos, publications

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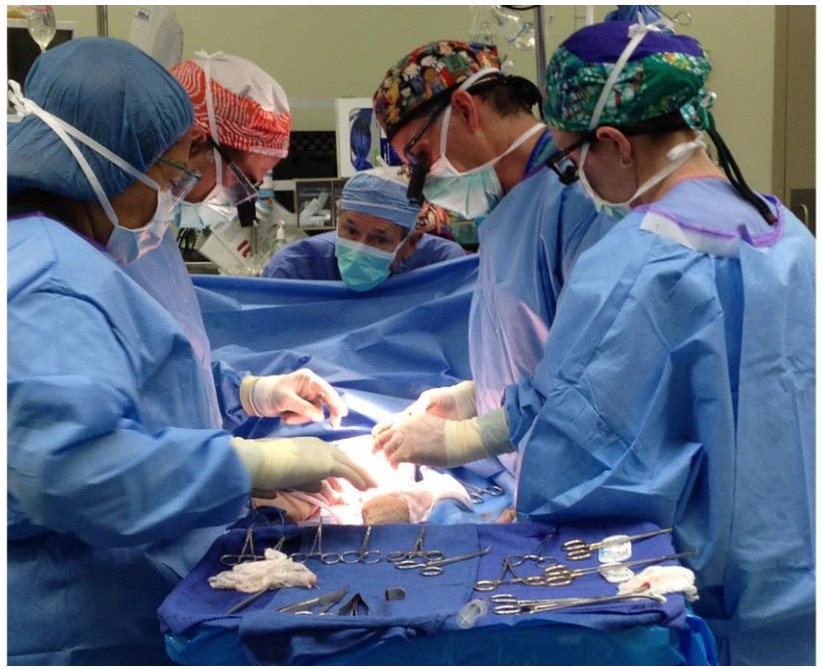
Collaboration

- *"It is the long history of humankind (and animal kind, too) that those who learned to collaborate and improvise most effectively have prevailed."*

Charles Darwin



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Collaboration at it's finest!

This image is actually 5 women decorated by world champion body-painter **Johannes Stotter**

MIBEC
Multi Institutional Bladder Exstrophy Consortium

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MIBEC
Multi Institutional Bladder Exstrophy Consortium

- **Boston Children's Hospital**
  - Dr. Joseph Borer
  - Dr. Rich Lee
  - Allysia Vesna, Research
- **Children's Hospital of Philadelphia**
  - Dr. Doug Canning
  - Dr. Aseem Shukla
  - Dr. Dana Weiss
  - Brynne Bonabitabo, NP
  - Jennifer Frazier, Research
- **Children's Wisconsin**
  - Dr. Mike Mitchell
  - Dr. John Kryger
  - Dr. Travis Groth
  - Dr. Elizabeth Roth
  - Coleen Rosen, DNP
  - Katie Sheridan, Research

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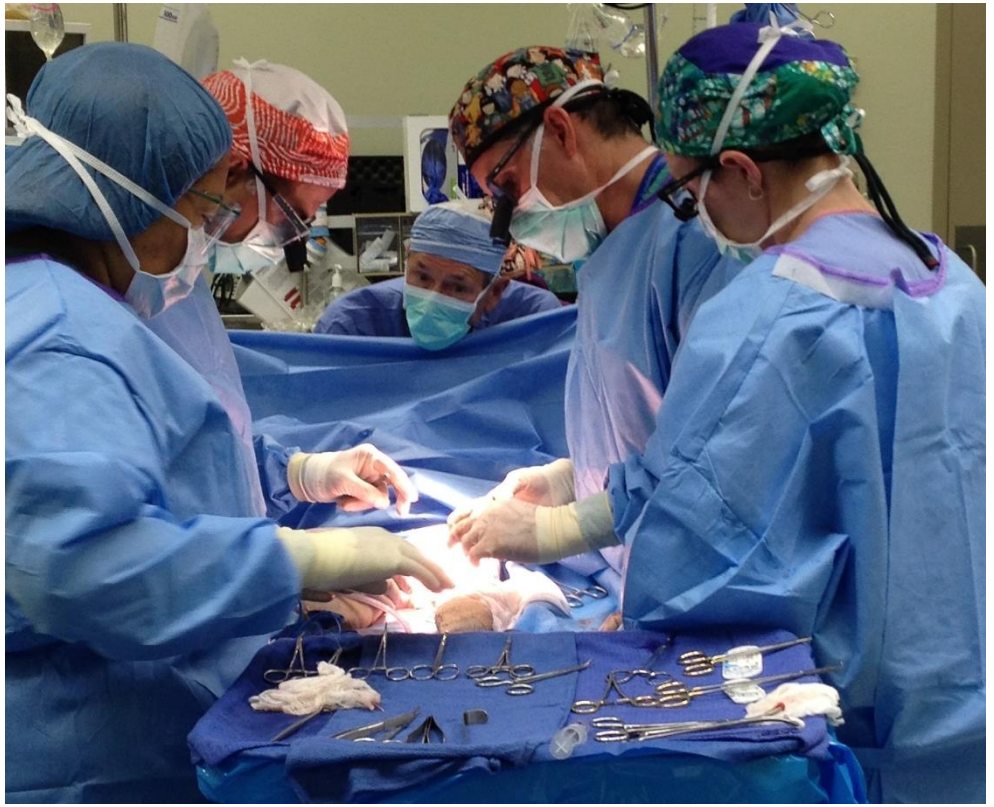
Insert Video of complete repair of bladder exstrophy



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Amy Wagner, MD

- Pediatric surgeon at Children's Wisconsin since 2009
- Professor at the Medical College of Wisconsin
- Earned her medical degree at Medical College of Wisconsin
- Completed her pediatric surgery fellowship at the University of California – San Francisco and Medical College of Wisconsin
- Dr. Wagner sees patients at the Milwaukee Hospital Campus and Mequon Clinic

Equipoise and Status of the Gastroschisis Outcomes of Delivery (GOOD) Study

Amy Wagner, MD
Professor of Surgery
Division of Pediatric Surgery
Fetal Concerns Center of Wisconsin

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GOOD
Gastroschisis Outcomes of Delivery

Disclosures

- No relevant financial relationships to disclose.

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GOOD
Gastroschisis Outcomes of Delivery

Background: Gastroschisis - Risk of Fetal Demise

Meta-analysis 2013

- 3276 pregnancies
  with gastroschisis

Prevalence **4.48/100**

- 7 times increased risk

RESEARCH www.AJOG.org

OBSTETRICS

Metaanalysis of the prevalence of intrauterine fetal death in gastroschisis

Andrew P. South, MD, MPH; Kevin M. Stutey, MD; Jareen Meinzen-Derr, PhD, MPH

OBJECTIVE: The objective of this study was to review the medical literature that has reported the risk for intrauterine fetal death (IUFD) in pregnancies with gastroschisis.

STUDY DESIGN: We systematically searched the literature to identify all published studies of IUFD and gastroschisis through June 2011 that were archived in MEDLINE, PubMed, or referenced in published manuscripts. The MESH terms *gastroschisis* or *abdominal wall defect* were used.

RESULTS: Fifty-four articles were included in the metaanalysis. There were 3276 pregnancies in the study and a pooled prevalence of IUFD of 4.48 per 100. Those articles that included gestational age of IUFD had a pooled prevalence of IUFD of 1.28 per 100 births at ≥36 weeks' gestation. The prevalence did not appear to increase at >35 weeks' gestation.

CONCLUSION: The overall incidence of IUFD in gastroschisis is much lower than previously reported. The largest risk of IUFD occurs before routine and elective early delivery would be acceptable. Risk for IUFD should not be the primary indication for routine elective preterm delivery in pregnancies that are affected by gastroschisis.

Key words: abdominal wall defect, fetal death, gastroschisis, IUFD, stillbirth

Cite this article as: South AP, Stutey KM, Meinzen-Derr J. Metaanalysis of the prevalence of intrauterine fetal death in gastroschisis. Am J Obstet Gynecol 2013;209:114.e1-13.

Gastroschisis is an abdominal wall defect of unclear cause and increasing incidence worldwide; current estimates are near 5 per 10,000 births.[1] There have been great improvements in survival in this patient population

★ EDITORS' CHOICE ★

because >95% of infants survive from birth to initial hospital discharge.[2] However, there remain many questions about perinatal management and, in particular, about the optimal gestational age at delivery. Intrauterine fetal death (IUFD) is more common in pregnancies that are affected by congenital anomalies. Among all major congenital anomalies, 2% of pregnancies result in stillbirth,[3] which is much higher than the 0.6% baseline rate in the general population.[4,5] This higher risk of stillbirth results in a higher frequency and level of antenatal monitoring and, in some cases, elective delivery at <39 weeks' gestation.[6] Decisions regarding obstetric management must be based on accurate knowledge of the risk for fetal death.

The mean age of spontaneous labor in pregnancies that are affected by gastroschisis is between 36 and 37 weeks' gestation,[7] yet the average age of delivery is approximately 1 week earlier. This discrepancy leads to the conclusion that infants with gastroschisis deliver early either for fetal/maternal indications or electively.[8] Although some clinicians advocate for early delivery to improve postnatal clinical outcomes (such as earlier initiation of enteral feeds and shorter hospitalization time), the literature does not document a consistent benefit.[9-11] Therefore, the primary rationale for elective delivery before the onset of labor may be the prevention of IUFD.[12]

The reported incidence of IUFD in pregnancies that are affected by gastroschisis is as high as 12.5%.[13] Although the cause for the increased risk of IUFD is unknown, hypotheses include umbilical cord compression after acute intestinal dilation,[14] oligohydramnios,[15] cardiovascular compromise that is related to high protein loss through the defect and subsequent hypovolemia,[16] and cytokine-mediated inflammation.[17,18] Additionally, there is increased risk for volvulus and vascular compromise that could lead to fetal death.[19] Studies that have documented high rates of IUFD are limited by small numbers, and many were conducted at a time when prenatal diagnosis of gastroschisis was uncommon. These studies found that most IUFDs occurred late in the third trimester. Obstetricians developed the practice of early elective delivery based on these studies. Additional

From the Divisions of Neonatology (Drs South and Stutey) and Biostatistics and Epidemiology (Dr Meinzen-Derr), Department of Pediatrics, Cincinnati Children's Hospital Medical Center, University of Cincinnati College of Medicine, Cincinnati, OH.

Received Jan. 30, 2013; revised April 2, 2013; accepted April 24, 2013.

This study was supported by the Perinatal Institute at Cincinnati Children's Hospital Medical Center, Cincinnati, OH.

The authors report no conflict of interest.

Presented at the annual meeting of the Pediatric Academic Societies, Boston, MA, April 28-May 1, 2012.

Reprints: Andrew P. South, MD, MPH, 3333 Burnet Ave., Cincinnati, OH 45229. andrew.south@cchmc.org.

0002-9378/free

http://dx.doi.org/10.1016/j.ajog.2013.04.032

For Editors' Commentary, see Contents

Source: Metaanalysis of the prevalence of intrauterine fetal death in gastroschisis. South AP, Stutey KM, Meinzen-Derr J Am J Obstet Gynecol. 2013;209-22

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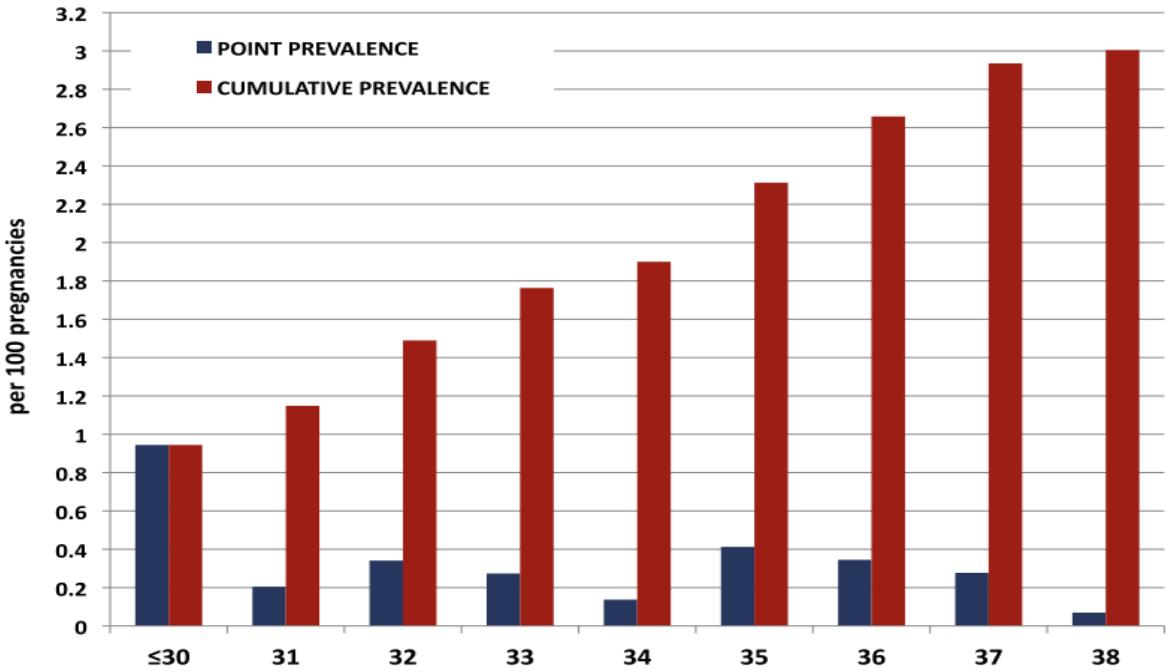
GOOD Gastroschisis Outcomes of Delivery

Background: Gastroschisis - Risk of Fetal Demise

- Pooled prevalence of IUFD of 1.28 per 100 births after 36 weeks
- Prevalence does not appear to increase after 35 weeks

FIGURE 3
Prevalence of intrauterine fetal death by gestational age



Cumulative and weekly prevalence of intrauterine fetal death per 100 pregnancies. *Blue lines* indicate weekly prevalence of intrauterine fetal death; *red lines* indicate cumulative prevalence of intrauterine fetal death.

South. Risk of IUFD in gastroschisis. Am J Obstet Gynecol 2013.

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GOOD
Gastroschisis Outcomes of Delivery

Background: Gastroschisis - Risk of Fetal Demise

US birth/death certificate data

- Retrospective cohort
- 860 cases of gastroschisis
- Stillbirth rate 4.8%

Source Sparks TN, Shaffer BL, Page J, et al. Gastroschisis: mortality risks with each additional week of expectant management. Am J Obstet Gynecol 2017;216:66.e1-7.

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OBSTETRICS

Gastroschisis: mortality risks with each additional week of expectant management

Teresa N. Sparks, MD; Brian L. Shaffer, MD; Jessica Page, MD; Aaron B. Caughey, MD, PhD

BACKGROUND: Prior studies have evaluated the overall risk of stillbirth in pregnancies with fetal gastroschisis. However, the gestational age at which mortality is minimized, balancing the risk of stillbirth against neonatal mortality, remains unclear.

OBJECTIVE: We sought to evaluate the gestational age at which prenatal and postnatal mortality risk is minimized for fetuses with gastroschisis.

STUDY DESIGN: This was a retrospective cohort study of singleton pregnancies delivered between 24 0/7 and 39 6/7 weeks, using 2005 through 2006 US national linked birth and death certificate data. Among pregnancies with fetal gastroschisis, prospective risk of stillbirth and risk of infant death were determined for each gestational age week. Risk of infant death with delivery was further compared to composite fetal/infant mortality risk with expectant management for 1 additional week.

RESULTS: Among 2,119,049 pregnancies, 860 cases (0.04%) of gastroschisis were identified. The overall stillbirth rate among gastroschisis cases was 4.8%, and infant death occurred in 8.3%. Prospective risk of stillbirth became more consistently elevated beginning at 35 weeks, rising to 13.9 per 1000 pregnancies (95% confidence interval, 10.8–17.1) at 39 weeks. Risk of infant death concurrently nadired in the third trimester, ranging between 62.4-66.8 per 1000 live births between 32-39 weeks. Comparing mortality with expectant management vs delivery, relative risk was significantly greater with expectant management between 37-39 weeks, reaching 1.90 (95% confidence interval, 1.73–2.08) at 39 weeks with a number needed to deliver of 17.49 (95% confidence interval, 15.34–20.32) to avoid 1 excess death.

CONCLUSION: Risk of prenatal and postnatal mortality for fetuses with gastroschisis may be minimized with delivery as early as 37 weeks.

Key words: gastroschisis, intrauterine fetal demise, neonatal death, stillbirth

Introduction

Gastroschisis occurs in approximately 3-5 of every 10,000 live births, with an estimated 90-97% of cases identified prenatally.[1,2] Most cases are thought to occur either sporadically or in a multifactorial nature, and risk factors include young maternal age, non-Hispanic white maternal race/ethnicity, tobacco use, and several environmental factors and medications.[1-4] The majority of gastroschisis cases occur as isolated defects, although 6-15% are associated with additional anomalies.[5-9] Important perinatal risks associated with gastroschisis include a 28% to >50% chance of early delivery, numerous concomitant morbidities, and mortality in up to 10% of infants.[1,3,8-10]

Several studies have evaluated stillbirth among cases of fetal gastroschisis, with overall rates reported from 0-14%.[1,6,7,9-12] Stillbirth risk may further increase in cases with concomitant oligohydramnios or when additional anomalies are present.[1,5] Many of the existing studies are based on small cohorts, though, and there has been limited investigation of stillbirth risk by gestational age (GA). A recent meta-analysis[12] found that stillbirth risk with fetal gastroschisis was greatest <36 weeks of gestation, and other literature has supported higher stillbirth rates throughout the third trimester in general.[1,13] Some studies have advocated for delivery ≤37 weeks of gestation because of increased stillbirth risk over time, potentially from increasing bowel inflammation and other factors.[14,15] Others have supported delivery at later term GAs due to improved neonatal outcomes and lower medical costs.[16,17] The GA at which mortality is minimized for fetuses with gastroschisis, balancing the risk of stillbirth against neonatal mortality, thus remains unclear.

We designed a large retrospective cohort study to investigate the prospective risk of stillbirth for each GA week among pregnancies with fetal gastroschisis, as well as the risk of neonatal death. Our objective was to determine the GA at which risk of mortality with expectant management begins to outweigh that with delivery of fetuses with gastroschisis, to improve our understanding of the GA at which mortality is minimized with delivery.

Materials and Methods

This was a retrospective cohort study of pregnancies in the United States during the years 2005 through 2006, using linked live birth certificate and infant death data from the National Center for Health Statistics (NCHS), Centers for Disease Control and Prevention (CDC).[18] Institutional review board approval and informed consent were not required for this study because of the publicly available and de-identified nature of the NCHS linked file.[19]

The NCHS established a national linked file using unique linkage numbers to match data from individual states. This file includes birth information for infants born in 2005 through 2006, as well as information about death if this occurred within the first 12 months of life (whether the death occurred in 2005, 2006, or 2007). A revised US Standard Certificate of Live Birth was implemented beginning in 2003, and was

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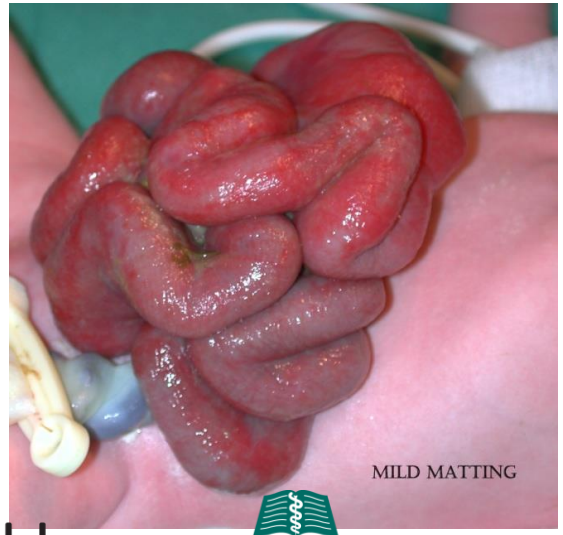
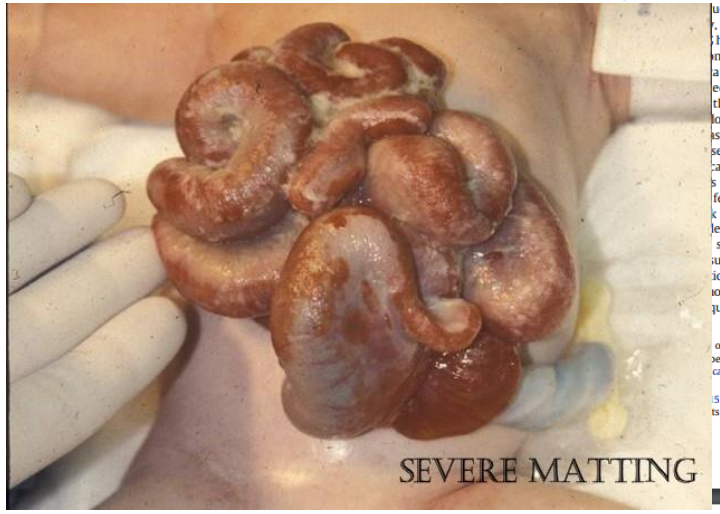
GOOD Gastroschisis Outcomes of Delivery

Gastroschisis: Bowel Morbidity

- CAPSNet prospective cohort study
- 692 patients
  - Validated GPS





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The correlation between the time spent in utero and the severity of bowel matting in newborns with gastroschisis

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ABSTRACT

Background: Optimal timing of delivery in fetuses with gastroschisis (GS) is unknown. Some favor early induced delivery to prevent bowel injury. This study evaluates the correlation between bowel injury and the gestational age at birth using the Gastroschisis Prognostic Score (GPS).
Methods: A national database was analyzed from 2005 to 2013. Patients were pooled based on their gestational age at birth. The mean GPS and % of patients with severe bowel matting were tabulated for each week in utero. Regression modeling was used to evaluate the relationship between the dependent (severe matting and GPS) and independent (gestational age) variables and the R^2 coefficient of determination was derived to evaluate model strength. Additional factors influencing the timing of delivery were evaluated.
Results: Of 780 cases, 88 were excluded because of missing data. A linear relationship is seen between increasing gestational age and decreasing bowel matting ($R^2 = 0.66$) and GPS ($R^2 = 0.72$). For every week in utero, the % of patients with severe matting decreases by 3.6%.
Conclusion: Early induced delivery simply to protect the bowel from ongoing in utero damage appears unfounded and should be reserved for evidence of closing gastroschisis or traditional obstetrical/fetal indications.

Gastroschisis (GS) is one of the most common congenital abdominal wall defects and is usually detected in the prenatal period in developed countries during routine fetal ultrasound and selective maternal serum
luent intrauterine growth restriction
, most babies with GS survive with an
have severe bowel damage and require
n [4]. The exact cause of bowel injury
auses include chemical irritation by
ed digestive enzymes in the amniotic
the restriction of venous and lymphatic
dominal wall defect [5–8]. A few authors
s the cause of bowel edema and have
section (CS) as a consequence [9–12].
cated for preterm delivery to prevent
s including ongoing bowel damage in
fetal demise, fetal distress with neuro-
k of "closing gastroschisis" leading to
lective preterm delivery has been asso-
stay, faster initiation to oral feeding
successful primary repair, and a shorter
ion [9,17]. On the other hand, preterm
own to increase the duration of hospi-
quired to reach full oral feeding [18–20].

In addition, premature GS babies are at higher risk of developing sepsis and cholestasis [4,21,22]. Many studies have been underpowered to detect significant outcome differences based on delivery practices, including the only prospective randomized trial published to date [23–25]. Additional limitations of the salient literature include the prolonged time span included in most studies resulting in the comparison of patients treated in different eras, inclusion of emergency and elective deliveries as well as the evaluation of patients without stratification based on disease severity.

In order to assess whether preterm delivery protects the bowel from ongoing damage in utero, the Canadian Pediatric Surgery Network (CAPSNet) database was used to correlate the time spent in utero with the severity of bowel matting and Gastroschisis Prognostic Score (GPS) in newborns with gastroschisis [26]. The study hypothesis was that if the proponents of preterm delivery were correct, we should see a higher percentage of severe matting and high-risk GPS with increasing gestational age.

1. Methods

1.1. Study population

The Canadian Pediatric Surgery Network includes all tertiary care
Canadian perinatal centers and has collected data on all congenit
diaphragmatic hernia and GS cases from fetal diagnosis until hosp
discharge or death since May 2005. CAPSNet is nested within a nati
universal health care delivery plan without appreciable private maternal

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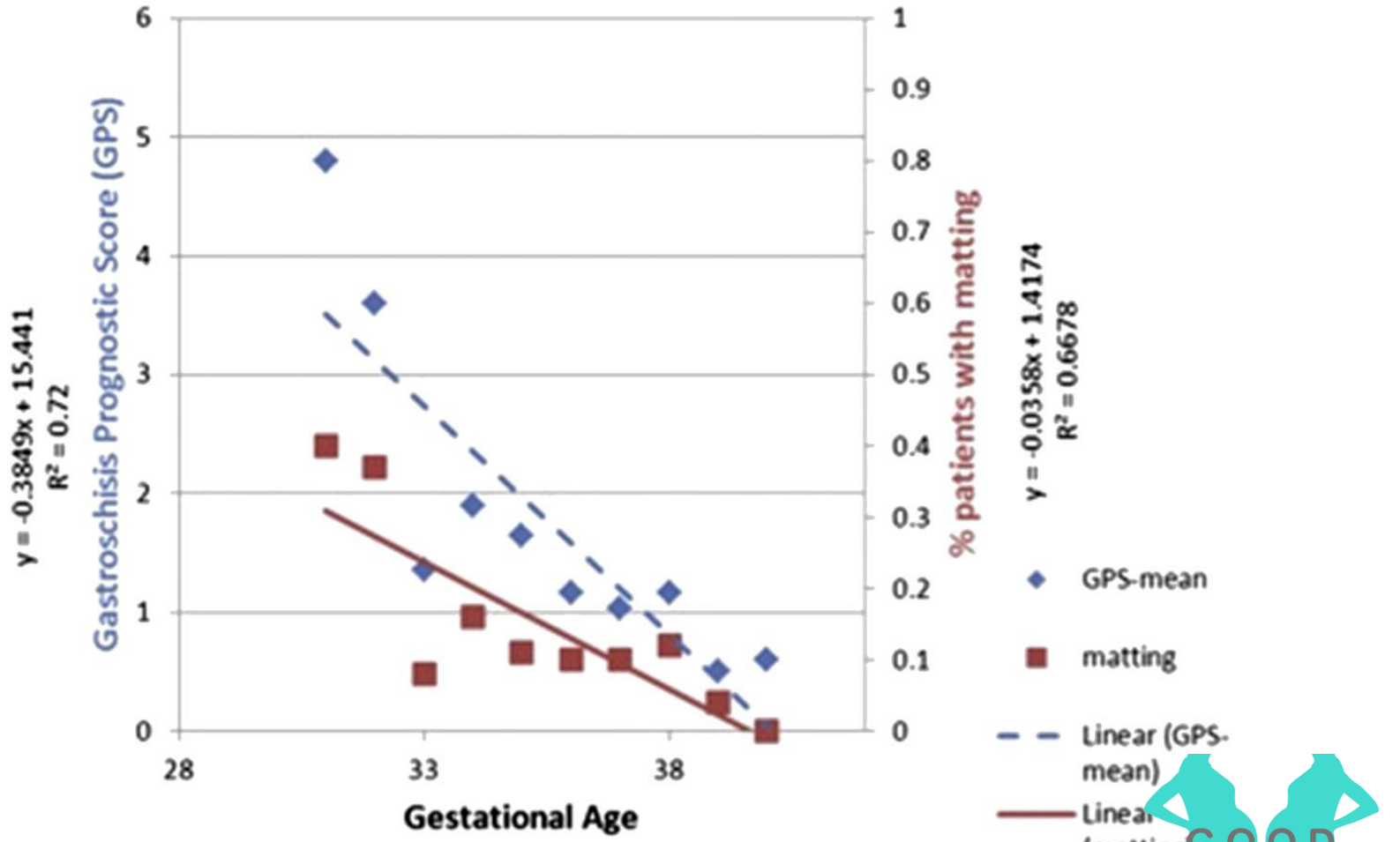
GOOD Gastroschisis Outcomes of Delivery

Gastroschisis: Bowel Morbidity

- Linear relationship found between increasing gestational age and *decreasing* bowel morbidity



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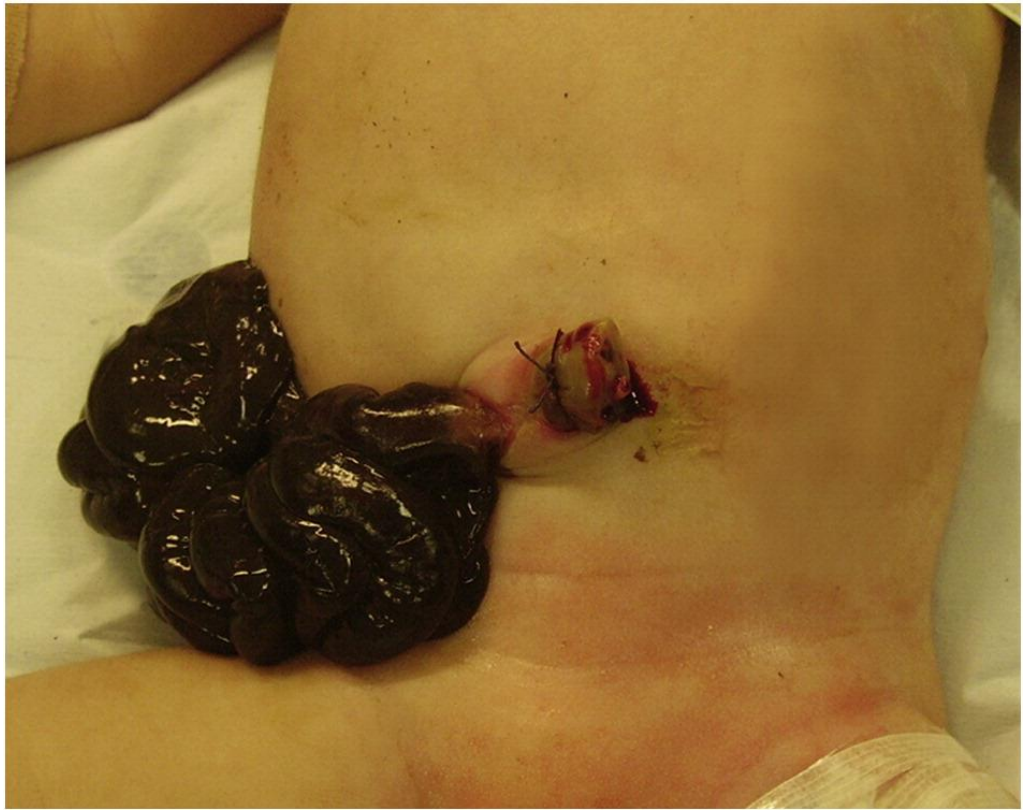
GOOD Gastroschisis Outcomes of Delivery

Gastroschisis: Bowel Morbidity

- Short gut syndrome
- Vanishing gastroschisis



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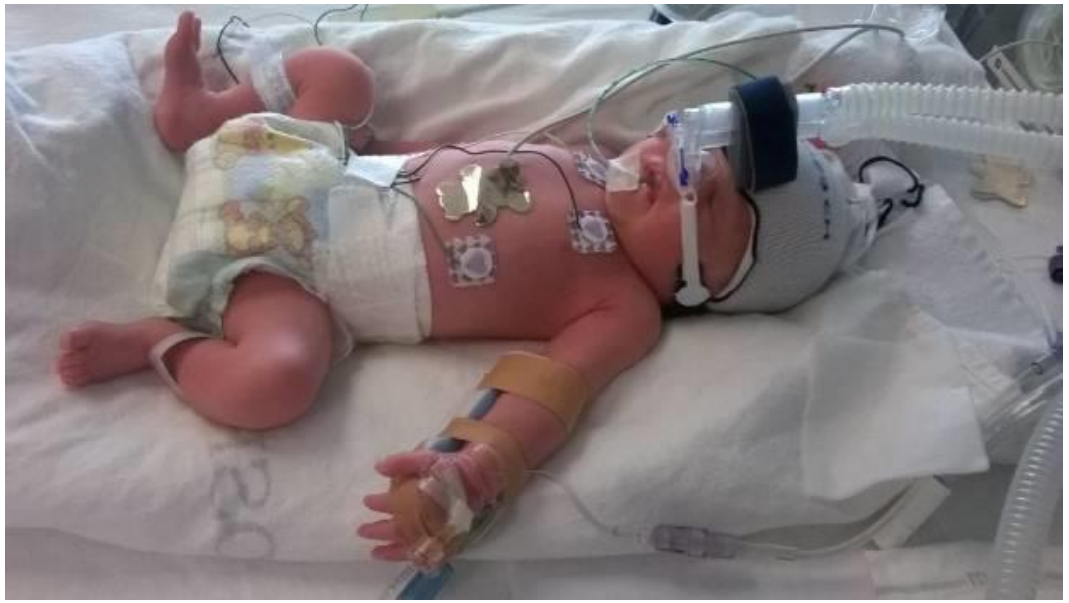
Early delivery?

- Preterm birth =
  - Respiratory morbidity
  - IVH
  - Sepsis
  - Need for phototherapy
  - Cognitive and motor developmental delays



Source: South AP, Marshall DD, Bose CL, Laughon MM. Growth and neurodevelopment at 16 to 24 months of age for infants born with gastroschisis. Journal of Perinatology. 2008;28(10):702-6.

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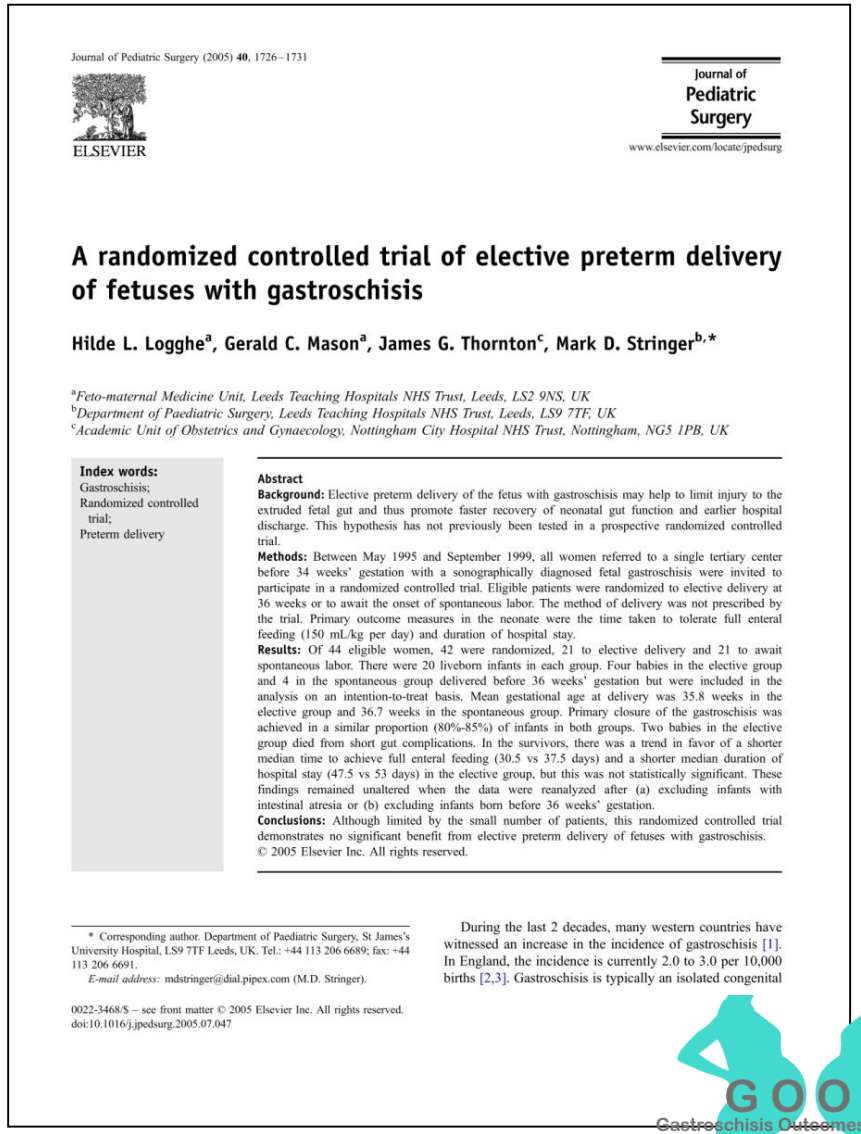
Early delivery?

Only previous RCT completed

- 42 patients randomized into two groups:
  - Spontaneous labor group (21) and an elective delivery at 36 weeks gestation group.
  - The elective delivery group trended toward better outcomes
    - Shorter time to full enteral feeds (30.5 vs. 37.5 days)
    - Shorter hospital LOS (47.5 days vs. 53 days)



Journal of Pediatric Surgery (2005) 40, 1726–1731

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Journal of Pediatric Surgery

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A randomized controlled trial of elective preterm delivery of fetuses with gastroschisis

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Index words:
Gastroschisis;
Randomized controlled trial;
Preterm delivery

Abstract

Background: Elective preterm delivery of the fetus with gastroschisis may help to limit injury to the extruded fetal gut and thus promote faster recovery of neonatal gut function and earlier hospital discharge. This hypothesis has not previously been tested in a prospective randomized controlled trial.

Methods: Between May 1995 and September 1999, all women referred to a single tertiary center before 34 weeks' gestation with a sonographically diagnosed fetal gastroschisis were invited to participate in a randomized controlled trial. Eligible patients were randomized to elective delivery at 36 weeks or to await the onset of spontaneous labor. The method of delivery was not prescribed by the trial. Primary outcome measures in the neonate were the time taken to tolerate full enteral feeding (150 mL/kg per day) and duration of hospital stay.

Results: Of 44 eligible women, 42 were randomized, 21 to elective delivery and 21 to await spontaneous labor. There were 20 liveborn infants in each group. Four babies in the elective group and 4 in the spontaneous group delivered before 36 weeks' gestation but were included in the analysis on an intention-to-treat basis. Mean gestational age at delivery was 35.8 weeks in the elective group and 36.7 weeks in the spontaneous group. Primary closure of the gastroschisis was achieved in a similar proportion (80%-85%) of infants in both groups. Two babies in the elective group died from short gut complications. In the survivors, there was a trend in favor of a shorter median time to achieve full enteral feeding (30.5 vs 37.5 days) and a shorter median duration of hospital stay (47.5 vs 53 days) in the elective group, but this was not statistically significant. These findings remained unaltered when the data were reanalyzed after (a) excluding infants with intestinal atresia or (b) excluding infants born before 36 weeks' gestation.

Conclusions: Although limited by the small number of patients, this randomized controlled trial demonstrates no significant benefit from elective preterm delivery of fetuses with gastroschisis.

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During the last 2 decades, many western countries have witnessed an increase in the incidence of gastroschisis [1]. In England, the incidence is currently 2.0 to 3.0 per 10,000 births [2,3]. Gastroschisis is typically an isolated congenital

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GOOD Gastroschisis Outcomes of Delivery

Source: Logghe HL, Mason GC, Thornton JG et al. J Pediatr Surg, 2005 Nov;40(11):1726-31.

Early delivery?

RCT attempted

- 34 wks versus routine care
- Primary outcome TPN duration
- 11 randomized to 34 week
- 10 to routine care
- Study stopped early because of increased sepsis in early group (40%
- (* major differences in study design)

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Elective delivery at 34 weeks *vs* routine obstetric care in fetal gastroschisis: randomized controlled trial

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KEYWORDS: early delivery; gastroschisis; neonatal sepsis; obstetric management; preterm delivery; randomized controlled trial

CONTRIBUTION

What are the novel findings of this work?
There is significantly more neonatal sepsis associated with early delivery of fetuses diagnosed with gastroschisis.

What are the clinical implications of this work?
Elective preterm delivery is detrimental to infants with gastroschisis.

to ED and 11 to RC. The trial was stopped at the first planned interim analysis due to patient safety concerns and for futility; thus, only 21 of the expected 86 patients (24.4%; 95% CI, 15.8–34.9%) were enrolled. Median gestational age at delivery was 34.3 (range, 34–36) weeks in the ED group and 36.7 (range, 27–38) weeks in the RC group. One patient in the ED group delivered at 36 weeks following unsuccessful induction at 34 weeks. Neonates of

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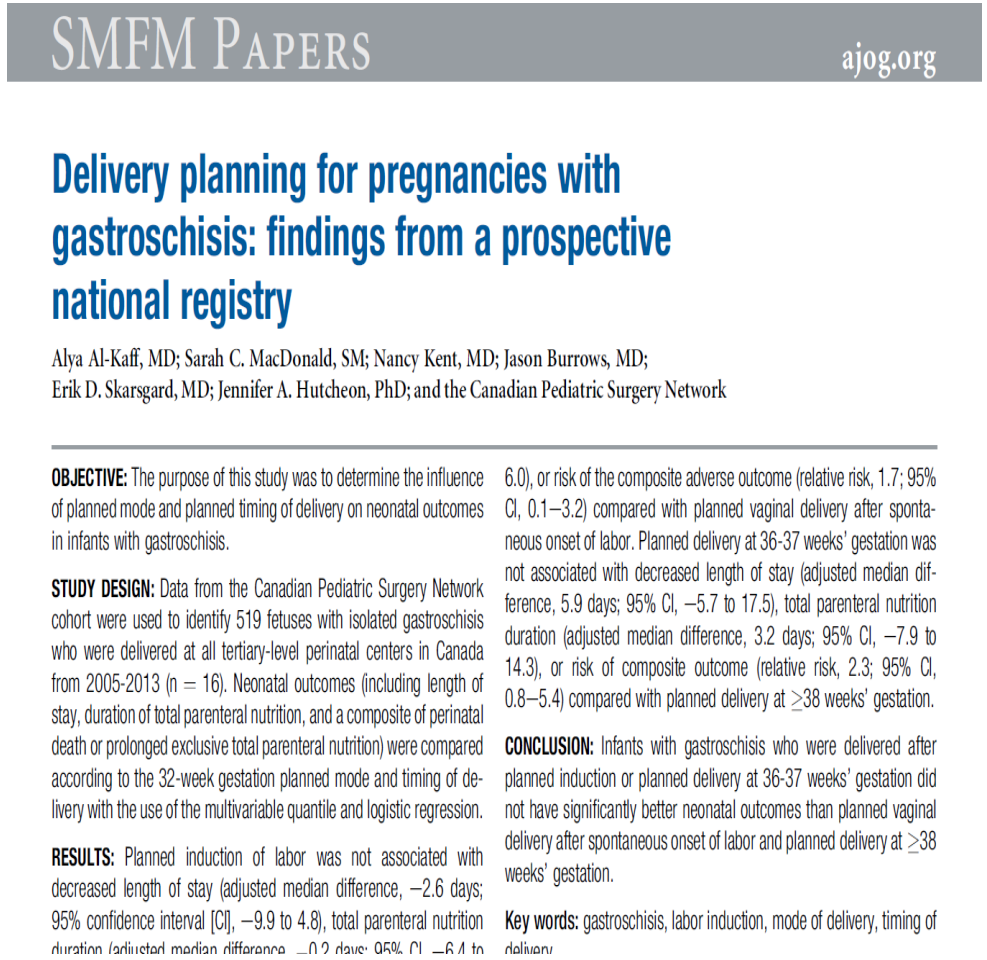
GOOD Gastroschisis Outcomes of Delivery

Source: Shamshirsaz AA, Lee TC, Hair AB, Erfani H, Espinoza J, Shamshirsaz AA, [illegible]KA, Ga[illegible]M, Nassr AA, Abrams SA, Mccullough LB, Chervenak FA, Olutoye OO, Belfort MA. Elective del[illegible]y at 34 [illegible]eks vs routine obstetric care in fetal gastroschisis: randomized controlled trial. Ultrasound Obstet Gynecol. 2020 Jan;55(1):15-19

Early delivery?

Prospective Study – Planned Delivery

- 519 babies CAPSNET prospective database
- Outcomes compared based on 32 week planned timing
- No difference in outcomes (LOS, TPN, enteral feeds, vent time, death) when planned 36-37 wk vs. ≥38
- Conclusion: “Nevertheless, a randomized trial would be needed to rule out potential for bias…”



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Delivery planning for pregnancies with gastroschisis: findings from a prospective national registry

Alya Al-Kaff, MD; Sarah C. MacDonald, SM; Nancy Kent, MD; Jason Burrows, MD; Erik D. Skarsgard, MD; Jennifer A. Hutcheon, PhD; and the Canadian Pediatric Surgery Network

OBJECTIVE: The purpose of this study was to determine the influence of planned mode and planned timing of delivery on neonatal outcomes in infants with gastroschisis.

STUDY DESIGN: Data from the Canadian Pediatric Surgery Network cohort were used to identify 519 fetuses with isolated gastroschisis who were delivered at all tertiary-level perinatal centers in Canada from 2005-2013 (n = 16). Neonatal outcomes (including length of stay, duration of total parenteral nutrition, and a composite of perinatal death or prolonged exclusive total parenteral nutrition) were compared according to the 32-week gestation planned mode and timing of delivery with the use of the multivariable quantile and logistic regression.

RESULTS: Planned induction of labor was not associated with decreased length of stay (adjusted median difference, −2.6 days; 95% confidence interval [CI], −9.9 to 4.8), total parenteral nutrition duration (adjusted median difference, −0.2 days; 95% CI, −6.4 to 6.0), or risk of the composite adverse outcome (relative risk, 1.7; 95% CI, 0.1–3.2) compared with planned vaginal delivery after spontaneous onset of labor. Planned delivery at 36-37 weeks' gestation was not associated with decreased length of stay (adjusted median difference, 5.9 days; 95% CI, −5.7 to 17.5), total parenteral nutrition duration (adjusted median difference, 3.2 days; 95% CI, −7.9 to 14.3), or risk of composite outcome (relative risk, 2.3; 95% CI, 0.8–5.4) compared with planned delivery at ≥38 weeks' gestation.

CONCLUSION: Infants with gastroschisis who were delivered after planned induction or planned delivery at 36-37 weeks' gestation did not have significantly better neonatal outcomes than planned vaginal delivery after spontaneous onset of labor and planned delivery at ≥38 weeks' gestation.

Key words: gastroschisis, labor induction, mode of delivery, timing of delivery

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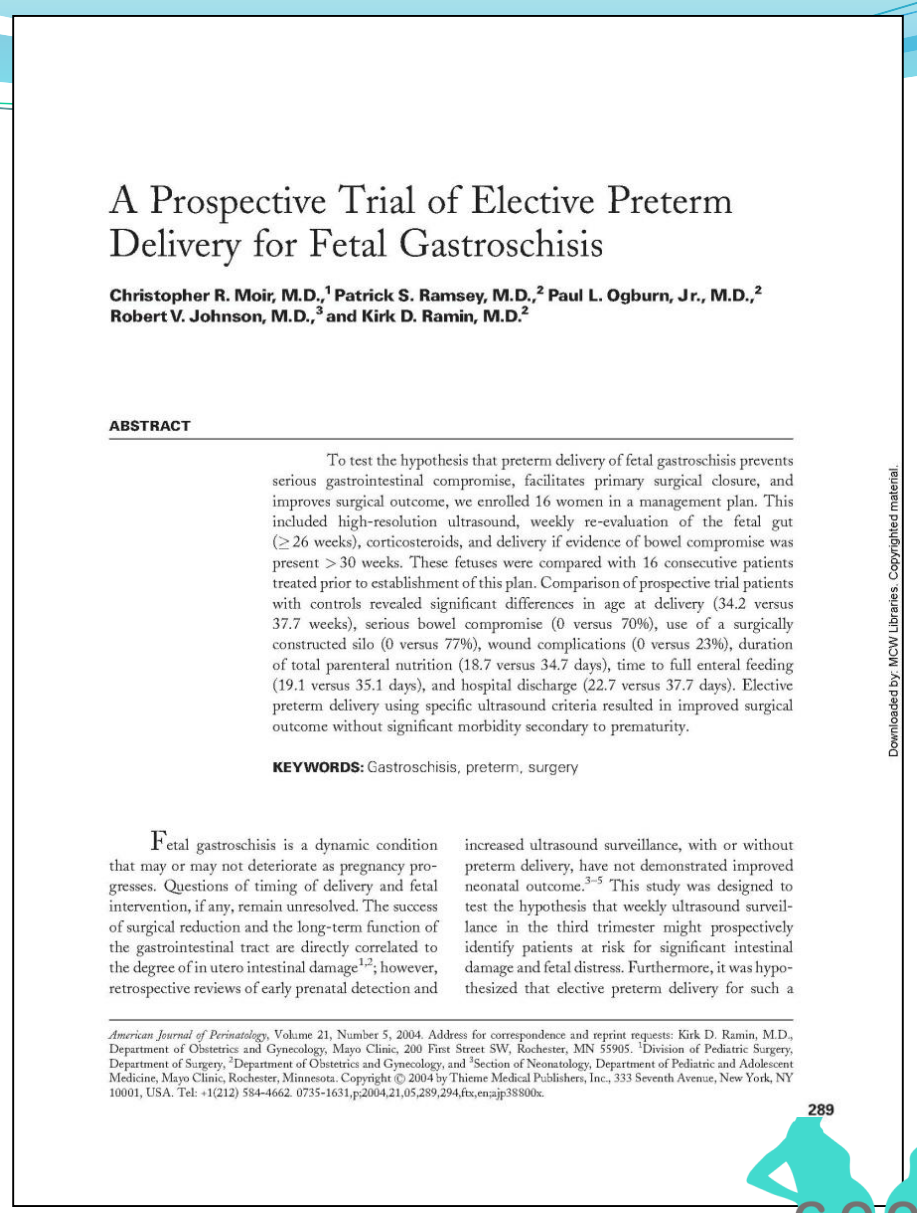
Al-Kaff A, MacDonald SC, Kent N, et al. Delivery planning for pregnancies with gastroschisis: findings from a prospective national registry. Am J Obstet Gynecol 2015;213:557.e1-8

Early delivery?

Prospective Study

- **32 patients in prospective trial**
  - **16 patients with expectant managment**
  - **16 patients after "mgmt plan"**
    - **Weekly US >26 wks**
    - **Delivery if concern for bowel compromise**
- **34.2 wks vs. 37.7 wks**
- Trend toward decreased TPN, time to enteral feeds, and LOS in early group



A Prospective Trial of Elective Preterm Delivery for Fetal Gastroschisis

Christopher R. Moir, M.D.,[1] Patrick S. Ramsey, M.D.,[2] Paul L. Ogburn, Jr., M.D.,[2] Robert V. Johnson, M.D.,[3] and Kirk D. Ramin, M.D.[2]

ABSTRACT

To test the hypothesis that preterm delivery of fetal gastroschisis prevents serious gastrointestinal compromise, facilitates primary surgical closure, and improves surgical outcome, we enrolled 16 women in a management plan. This included high-resolution ultrasound, weekly re-evaluation of the fetal gut (≥ 26 weeks), corticosteroids, and delivery if evidence of bowel compromise was present > 30 weeks. These fetuses were compared with 16 consecutive patients treated prior to establishment of this plan. Comparison of prospective trial patients with controls revealed significant differences in age at delivery (34.2 versus 37.7 weeks), serious bowel compromise (0 versus 70%), use of a surgically constructed silo (0 versus 77%), wound complications (0 versus 23%), duration of total parenteral nutrition (18.7 versus 34.7 days), time to full enteral feeding (19.1 versus 35.1 days), and hospital discharge (22.7 versus 37.7 days). Elective preterm delivery using specific ultrasound criteria resulted in improved surgical outcome without significant morbidity secondary to prematurity.

KEYWORDS: Gastroschisis, preterm, surgery

Fetal gastroschisis is a dynamic condition that may or may not deteriorate as pregnancy progresses. Questions of timing of delivery and fetal intervention, if any, remain unresolved. The success of surgical reduction and the long-term function of the gastrointestinal tract are directly correlated to the degree of in utero intestinal damage[1,2]; however, retrospective reviews of early prenatal detection and increased ultrasound surveillance, with or without preterm delivery, have not demonstrated improved neonatal outcome.[3–5] This study was designed to test the hypothesis that weekly ultrasound surveillance in the third trimester might prospectively identify patients at risk for significant intestinal damage and fetal distress. Furthermore, it was hypothesized that elective preterm delivery for such a

American Journal of Perinatology, Volume 21, Number 5, 2004. Address for correspondence and reprint requests: Kirk D. Ramin, M.D., Department of Obstetrics and Gynecology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905. [1]Division of Pediatric Surgery, Department of Surgery, [2]Department of Obstetrics and Gynecology, and [3]Section of Neonatology, Department of Pediatric and Adolescent Medicine, Mayo Clinic, Rochester, Minnesota. 0735-1631,p;2004,21,05,289,294,ftx,en;ajp38800x.

Moir CR, Ramsey PS, Ogburn PL et al. 2004
American Journal of Perinatology, 21:5,289-94.

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Gastroschisis Outcomes of Delivery

Early delivery?

"There is a lack of published data in this area," and "further trials are needed."

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GOOD Gastroschisis Outcomes of Delivery

Grant NH, Dorling J, Thornton JG. Elective preterm birth for fetal gastroschisis. Cochrane Database Syst Rev 2013.

Early delivery?

Meta-analysis 2017

- 1430 patients (18 studies) included
- Decreases in
  - Sepsis (p<0.01)
  - Less time to full feeds (p=0.03)
  - Less time on TPN (p=0.07) in pre-term cohort

Outcomes of gastroschisis early delivery: A systematic review and Meta-analysis. Landish RM, Yin Z, Christensen MA, Szabo A, Wagner AJ. J Pediatr Surg 2017(52:1)1962-71.

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Outcomes of gastroschisis early delivery: A systematic review and meta-analysis☆

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Delivery
Elective
Feeding
Preterm
Outcome

ABSTRACT

Background/purpose: Elective preterm delivery (EPD) of a fetus with gastroschisis may prevent demise and ameliorate intestinal injury. While the literature on optimal timing of delivery varies, we hypothesize that a potential benefit may be found with EPD.

Methods: A meta-analysis of publications describing timing of delivery in gastroschisis from 1/1990 to 8/2016 was performed, including studies where either elective preterm delivery (group 1, G1) or preterm gestational age (GA) (group 2, G2) were evaluated against respective comparators. The following outcomes were analyzed: total parenteral nutrition (TPN), first enteral feeding (FF), length of stay, ventilator days, fetal demise, complex gastroschisis, sepsis, and death.

Results: Eighteen studies describing 1430 gastroschisis patients were identified. G1 studies found less sepsis ($p < 0.01$), fewer days to FF ($p = 0.03$), and 11 days less of TPN ($p = 0.07$) in the preterm cohort. Comparatively, G2 studies showed less days to FF in term GA ($p = 0.02$).Whereas G1 BWs were similar, G2 preterm had a significantly lower BW compared to controls ($p = 0.001$).

Conclusions: Elective preterm delivery appears favorable with respect to feeding and sepsis. However, benefits are lost when age is used as a surrogate of EPD. A randomized, prospective, multi-institutional trial is necessary to delineate whether EPD is advantageous to neonates with gastroschisis.

Type of study: Treatment study.

Level of evidence: Level III.

Gastroschisis, a common congenital abdominal wall abnormality that causes the intestines of a fetus to herniate into the amniotic fluid, has an incidence of 1 in every 4000 births, and its prevalence is increasing [1, 2]. Pregnancies complicated by fetal gastroschisis have a 7-fold higher rate of fetal demise or stillbirth compared with normal pregnancies [3, 4]. Other gastroschisis-associated complications, such as bowel injury, may occur later in the pregnancy. Undoubtedly, neonatal gut dysfunction heightens the morbidity of gastroschisis newborns, and may result in total parenteral nutrition (TPN) requirements and prolonged hospital length of stay (LOS) [5]. As a result, many clinicians elect to deliver as early as 36 weeks in an attempt to minimize the risk of demise, caustic exposure to the intestines, and thereby mitigate gastroschisis morbidity and mortality.

However, early delivery has its own potential set of complications, including increased mortality, respiratory morbidity, cholestasis and cognitive deficits [6–8]. Currently, there is no consensus on the ideal timing of delivery in cases of fetal gastroschisis, resulting in practice variations; for this reason, data-driven conclusions are essential to understand the risks and benefits of elective preterm delivery. Although substantial efforts have been made over the past two decades to delineate such risks, the literature varies considerably with both study design and outcomes, making interpretation of data challenging. Therefore, the objective of this investigation was to compare feeding and neonatal outcomes of infants with gastroschisis who underwent preterm delivery to those who were expectantly managed or delivered at term through a formal systematic literature review and meta-analysis.

1. Materials and methods

1.1. Search strategy

A systematic review was undertaken following PRISMA [illegible] [9]. A systematic search of published literature was perfor[illegible] 2016 using the following sources: PubMed, MEDLI[illegible]SCO[illegible] Cochrane Library databases. Limited to English langua[illegible] noncase reports published between 1/1990 and 08/03/2016, th[illegible]

☆ There are no conflicts of interest. No funding was received.

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http://dx.doi.org/10.1016/j.jpedsurg.2017.08.068
0022-3468/

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Hypothesis

- Delivery at 35 0/7- 35 6/7 weeks in stable patients with gastroschisis is superior to observation and expectant management with a goal of delivery at 38 0/7 - 38 6/7 weeks.

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GOOD Study Principal Investigators

- **Anthony Johnson, DO** – University of Texas Health Center, Houston, TX (MFM)
- **Aniko Szabo, PhD** – Medical College of Wisconsin, Milwaukee, WI (Biostatistics)
- **Amy Wagner, MD** – Children's Wisconsin, Milwaukee, WI
- **Barbara Warner, MD** – Washington University, St. Louis, MO (Neonatology)

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Sub Committees

- **Maternal Fetal Medicine**
  - Alain Gagnon, MD
  - Anthony Johnson, DO
  - Richard O'Shaughnessy, MD
  - Erika Peterson, MD
  - Bill Polzin, MD
  - Britton Rink, MD
- **Neonatology**
  - Steve Leuthner, MD, MA
  - Barb Warner, MD
  - Heidi Karpen, MD
- **Pediatric Surgery**
  - Terry Buchmiller, MD
  - Brad Feltis, MD, PhD
  - Foong-Yen Lim, MD
  - Francois Luks, MD
  - Kuojen Tsao, MD
  - Amy Wagner, MD
- **Statistics**
  - Aniko Szabo, PhD

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Specific Aims

- **Specific Aim 1.** To determine the clinical risks of infants with gastroschisis who were delivered at 35 weeks compared with those of infants who were expectantly managed to observed delivery with a goal of 38 weeks.

- **Specific Aim 2.** To determine maternal and neonatal intermediate outcomes for cases of gastroschisis in the 35-week delivery group compared with the expectant management with a goal of 38 weeks group.

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Study Design

- Patients may be enrolled any time after diagnosis up to 33 6/7 weeks.
- Patients will be randomized at 33 weeks gestation.
  - Delivery at 35 0/7 weeks through 35 6/7 weeks.
  - Expectant management with induction at 38 0/7 through 38 6/7.
- With the exception of delivery timing, all medical care will be given based on the standard practices of each treating hospital.
- Minimum monitoring will be weekly BPP or NST with AF assessment, monthly EFW beginning at 32 weeks.
- (Management per local protocol with minimum monitoring)

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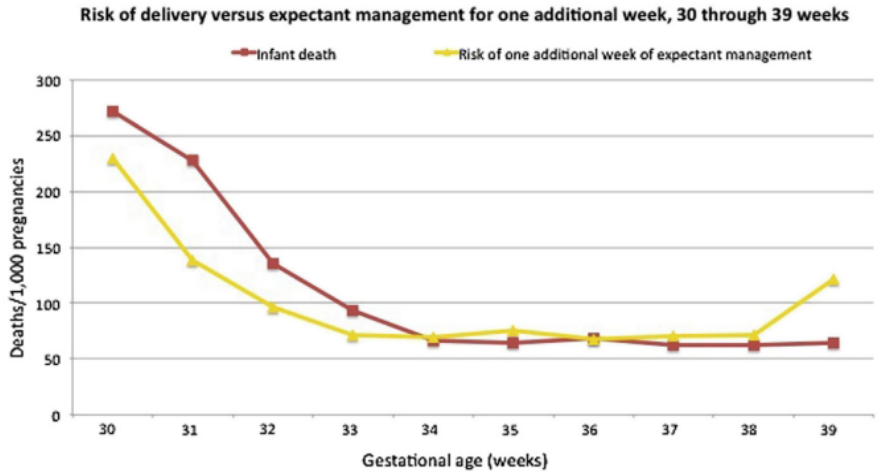
35 vs. 38 weeks

FIGURE
Mortality risk with delivery versus composite risk with expectant management



Composite risk of expectant management for 1 additional week is derived from the sum of the risk of stillbirth during each gestational age week plus risk of infant death with delivery during the subsequent week.

Sparks et al. Risk of mortality with gastroschisis. Am J Obstet Gynecol 2017.

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OBSTETRICS

Gastroschisis: mortality risks with each additional week of expectant management

Teresa N. Sparks, MD; Brian L. Shaffer, MD; Jessica Page, MD; Aaron B. Caughey, MD, PhD

BACKGROUND: Prior studies have evaluated the overall risk of stillbirth in pregnancies with fetal gastroschisis. However, the gestational age at which mortality is minimized, balancing the risk of stillbirth against neonatal mortality, remains unclear.
OBJECTIVE: We sought to evaluate the gestational age at which prenatal and postnatal mortality risk is minimized for fetuses with gastroschisis.
STUDY DESIGN: This was a retrospective cohort study of singleton pregnancies delivered between 24 0/7 and 39 6/7 weeks, using 2005 through 2006 US national linked birth and death certificate data. Among pregnancies with fetal gastroschisis, prospective risk of stillbirth and risk of infant death were determined for each gestational age week. Risk of infant death with delivery was further compared to composite fetal/infant mortality risk with expectant management for 1 additional week.
RESULTS: Among 2,119,049 pregnancies, 860 cases (0.04%) of gastroschisis were identified. The overall stillbirth rate among gastroschisis cases was 4.8%, and infant death occurred in 8.3%. Prospective risk of stillbirth became more consistently elevated beginning at 35 weeks, rising to 13.9 per 1000 pregnancies (95% confidence interval, 10.8—17.1) at 39 weeks. Risk of infant death concurrently nadired in the third trimester, ranging between 62.4-66.8 per 1000 live births between 32-39 weeks. Comparing mortality with expectant management vs delivery, relative risk was significantly greater with expectant management between 37-39 weeks, reaching 1.90 (95% confidence interval, 1.73—2.08) at 39 weeks with a number needed to deliver of 17.49 (95% confidence interval, 15.34—20.32) to avoid 1 excess death.
CONCLUSION: Risk of prenatal and postnatal mortality for fetuses with gastroschisis may be minimized with delivery as early as 37 weeks.

Key words: gastroschisis, intrauterine fetal demise, neonatal death, stillbirth

Introduction

Gastroschisis occurs in approximately 3-5 of every 10,000 live births, with an estimated 90-97% of cases identified prenatally.[1,2] Most cases are thought to occur either sporadically or in a multifactorial nature, and risk factors include young maternal age, non-Hispanic white maternal race/ethnicity, tobacco use, and several environmental factors and medications.[1-4] The majority of gastroschisis cases occur as isolated defects, although 6-15% are associated with additional anomalies.[5-9] Important perinatal risks associated with gastroschisis include a 28% to >50% chance of early delivery, numerous concomitant morbidities, and mortality in up to 10% of infants.[1,3,8-10]

Several studies have evaluated stillbirth among cases of fetal gastroschisis, with overall rates reported from 0-14%.[1,6,7,9-12] Stillbirth risk may further increase in cases with concomitant oligohydramnios or when additional anomalies are present.[1,5] Many of the existing studies are based on small cohorts, though, and there has been limited investigation of stillbirth risk by gestational age (GA). A recent meta-analysis[12] found that stillbirth risk with fetal gastroschisis was greatest <36 weeks of gestation, and other literature has supported higher stillbirth rates throughout the third trimester in general.[1,13] Some studies have advocated for delivery ≤37 weeks of gestation because of increased stillbirth risk over time, potentially from increasing bowel inflammation and other factors.[14,15] Others have supported delivery at later term GAs due to improved neonatal outcomes and lower medical costs.[16,17] The GA at which mortality is minimized for fetuses with gastroschisis, balancing the risk of stillbirth against neonatal mortality, thus remains unclear.

We designed a large retrospective cohort study to investigate the prospective risk of stillbirth for each GA week among pregnancies with fetal gastroschisis, as well as the risk of neonatal death. Our objective was to determine the GA at which risk of mortality with expectant management begins to outweigh that with delivery of fetuses with gastroschisis, to improve our understanding of the GA at which mortality is minimized with delivery.

Materials and Methods

This was a retrospective cohort study of pregnancies in the United States during the years 2005 through 2006, using linked live birth certificate and infant death data from the National Center for Health Statistics (NCHS), Centers for Disease Control and Prevention (CDC).[18] Institutional review board approval and informed consent were not required for this study because of the publicly available and de-identified nature of the NCHS linked file.[19]

The NCHS established a national linked file using unique linkage numbers to match data from individual states. This file includes birth information for infants born in 2005 through 2006, as well as information about death if this occurred within the fi[illegible] months of life (whether the deat[illegible] in 2005, 2006, or 2007). A[illegible]vis[illegible]Standard Certificate of Live[illegible] mplemented beginning in

Cite this article as: Sparks TN, Shaffer BL, Page J, et al. Gastroschisis: mortality risks with each additional week of expectant management. Am J Obstet Gynecol 2017;216:66.e1-7.

0002-9378/$36.00

http://dx.doi.org/10.1016/j.ajog.2016.08.036

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Sparks TN, Shaffer BL, Page J, et al. Gastroschisis: mortality risks with each additional week of expectant management. Am J Obstet Gynecol 2017;216:66.e1-7.

Inclusion Criteria:

- **A.)** English or Spanish speaking

  (open to other languages)
- **B.)** Age ≥18 years
- **C.)** Sonographic diagnosis of GS ≤33 weeks' gestation
- **D.)** Singleton pregnancy
- **E.)** Written informed consent for participation.

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Exclusion Criteria

- **A.)** Fetal anomaly unrelated to GS (e.g., chromosomal abnormality, congenital structural abnormality)
- **B.)** Severe intrauterine growth restriction – defined as growth below the 5th percentile for age
- **C.)** Previous history of stillbirth or preterm delivery (<36 weeks)
- **D.)** Maternal hypertension or insulin dependent diabetes
- **E.)** Prenatal care began after 24 weeks of pregnancy.

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Exclusion Criteria (con't):

- **F.)** Unstable pregnancy – defined as meeting any of the following
  - Amniotic fluid assessments with maximal vertical pocket <2cm or >8cm in third trimester
  - Umbilical artery Dopplers with S/D ratio or resistive index >97th percentile for age with or without absent or reverse end diastolic flow
  - Non-stress test and/ or biophysical profile deemed non-reassuring by treating clinician

- **G.)** Subject cannot consent on own behalf or not their own legally authorized representative

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Primary Composite Outcome

- **Stillbirth**
- **Neonatal Death Prior to Discharge**
- **Need for Parenteral Nutrition at 30 days**
  - Defined as being on PN (TPN or IL) at day 30
- **Respiratory Morbidity**
  - Defined as within the first 72 hours (exclusive of procedure):
    - Use of CPAP or high-flow N/C for at least 2 hours
    - Supplemental O2 of 0.3 for 4 hours
    - Mechanical ventilation
    - ECMO
- **Sepsis**

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Secondary Outcomes

- **Maternal**
  - Need for cesarean section
  - Spontaneous labor vs. required induction of labor
  - Chorioamnionitis
  - Need for transfusion
  - VTE

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Secondary Outcomes (con't)

- **Neonatal**
  - Birth weight
  - CVL days
  - Intestinal atresia
  - Sepsis
  - NEC
  - Need for caffeine
  - Need for surfactant
  - Length of stay
  - Discharge weight

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Secondary Outcomes (con't)

Surgical

- Date and location of closure
- Timing after birth of closure
- Side and Diameter of Defect
- Closure technique
- Anesthesia exposure

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Tertiary Outcomes

- Investigate the validity of the diagnosis of intrauterine growth restriction using the Hadlock, Shepard, and Siemer Fetal Weight (EFW)
- Investigate the validity of the CAPSNET Gastroschisis Prognostic Score to predict intestinal morbidity and time to full feeds
- Investigate the use of intraperitoneal pressure monitoring during closure to predict intestinal morbidity and complication rate

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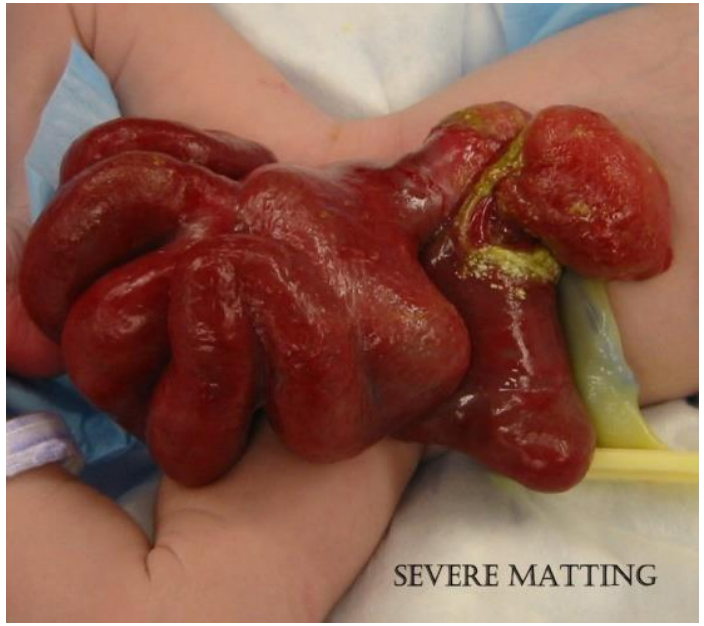
Tertiary Outcomes (cont'd)

Gastroschisis Prognostic Score

- Own prospective study verifying the usefulness of this score
- Scoring to be done at bedside by surgeon and/or by our specialist based on photographs
- Interested in comparing both scoring methods for validity and correlation to outcomes
- Dr. Buchmiller validating



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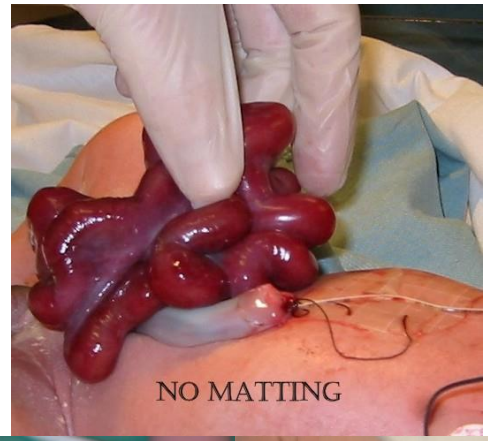
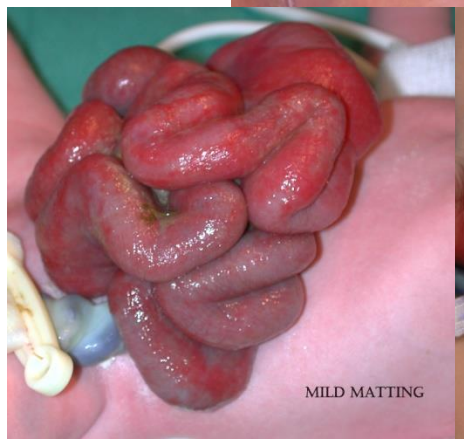
Tertiary Outcomes (cont'd)

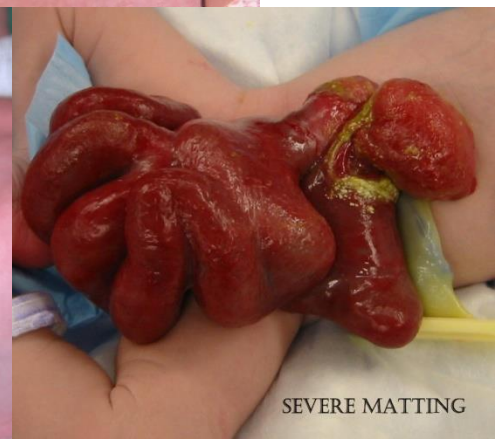
Bowel Injury Score

Bowel Matting	0 – None	1 – Mild	2 – Severe
Bowel Necrosis	0 – Absent	1 – Focal	2- Diffuse
Bowel Atresia	0 – Absent	1 – Suspected	2 – Present
Bowel Perforation	0 – Absent	2 – Present	

We will track date and time of photographs and bedside scoring.







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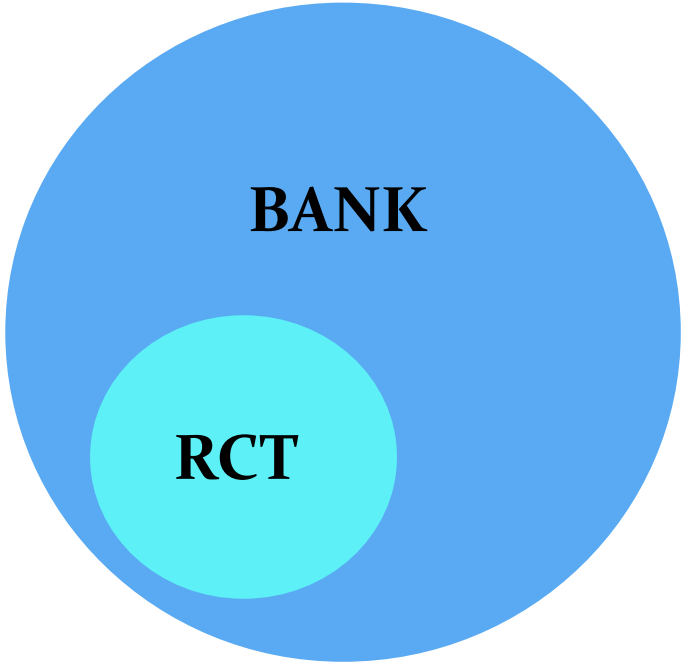
Two consents/Protocols

- RCT
- Data bank
  - Consent patients for continued prospective data collection up to 5 years
  - Further opportunity for long-term neurodevelopmental outcomes
  - Biobank (blood sample)



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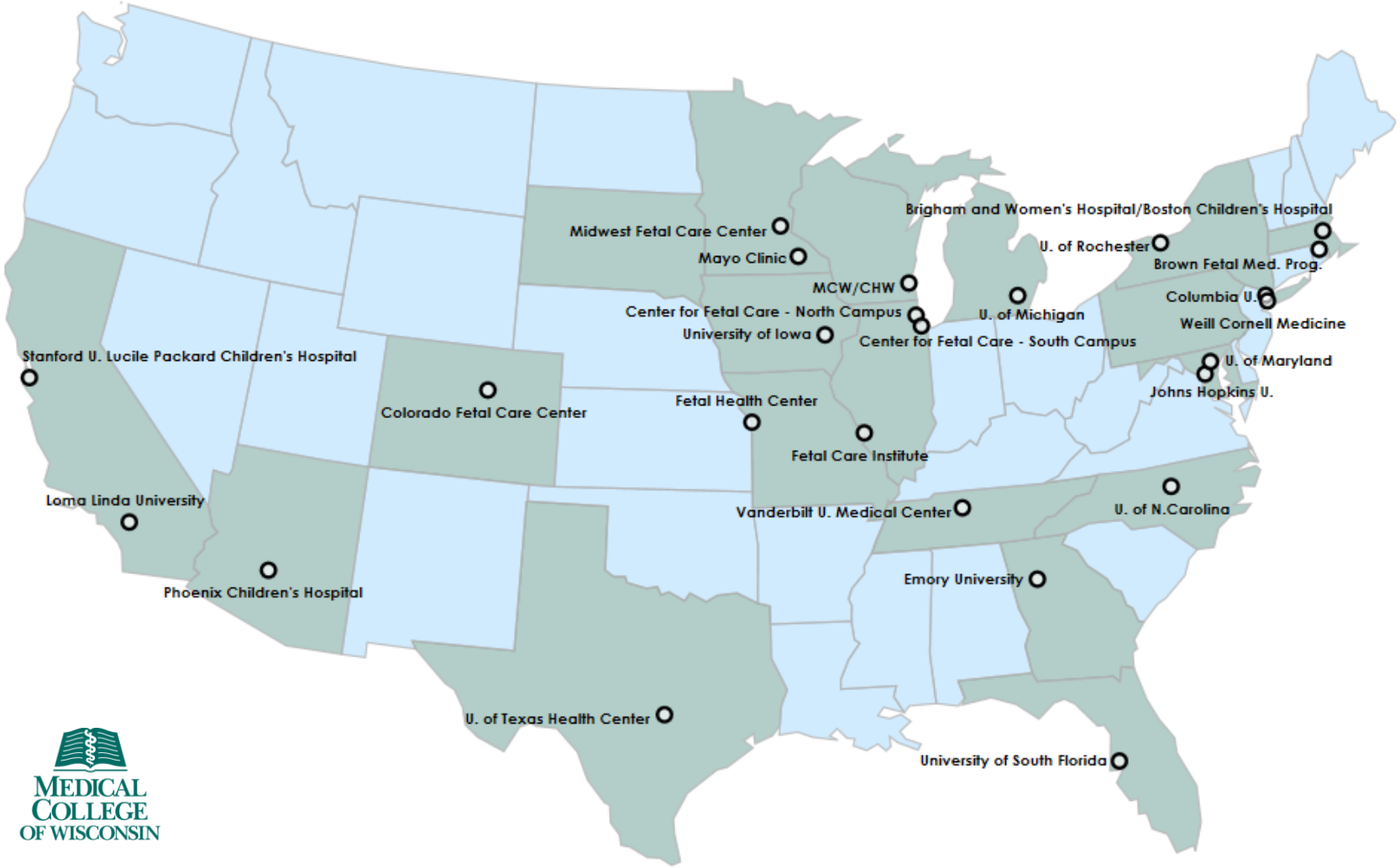
Study Status:



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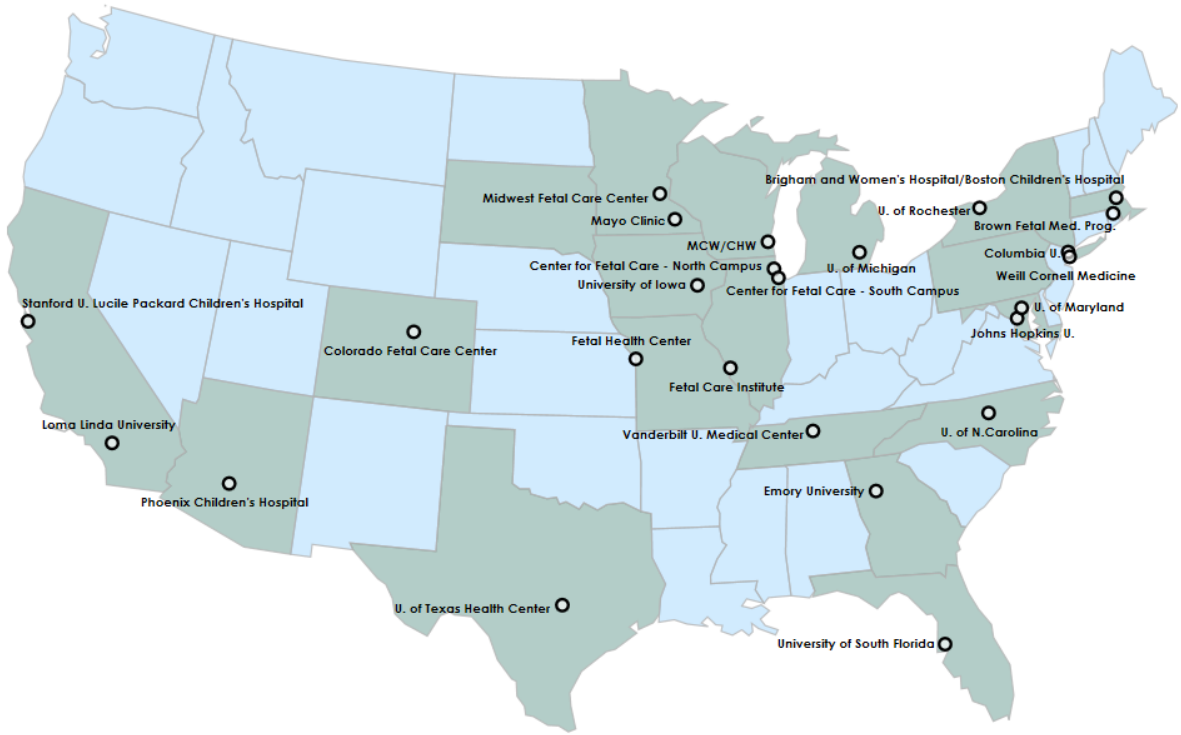
Study Status: (n = 800)

- Centers enrolling: 26
- Patients screened: 76
- Patients consented: 44
- Patients completed: 27



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Data Safety Monitoring Board:

- Assembled, met four times
- Includes each subspecialty:
  - Neonatology (Chair)
  - MFM
  - Pediatric Surgery
  - Biostatistics
  - Mediator

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Funding:

- Funded currently for **$1500 stipend** per patient enrollment
  - West Foundation Grant
  - WeCARE Grant
  - Pick Foundation Trust Grant
  - NIH **R01HD104607** Eunice Kennedy Shriver National Institute of Child Health & Human Development (NICHD)

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Next steps:

- R01 2023: Long-term Neurodevelopmental outcomes
  - Susan Cohen, MD

- R01 2023: Long-term GI/Motility outcomes
  - Katja Kovacic, MD

- R01 2024: Genetic/Epigenetic basis for gastroschisis
  - Honey Reddi, PhD (MCW PML)
  - Angie Jelin, MD

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Thank you!

awagner@chw.org

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David Saudek, MD

- Pediatric Cardiologist at Children's Wisconsin since 2007
- Fetal cardiologist at Herma Heart Institute
- Associate professor at the Medical College of Wisconsin
- Earned his medical degree from the Medical College of Georgia
- Completed his fellowship at Cleveland Clinic Foundation - Ohio and Medical College of Wisconsin – Milwaukee
- Dr. Saudek sees patients at the Milwaukee Hospital Campus, Neenah Clinic, and New Berlin Clinic

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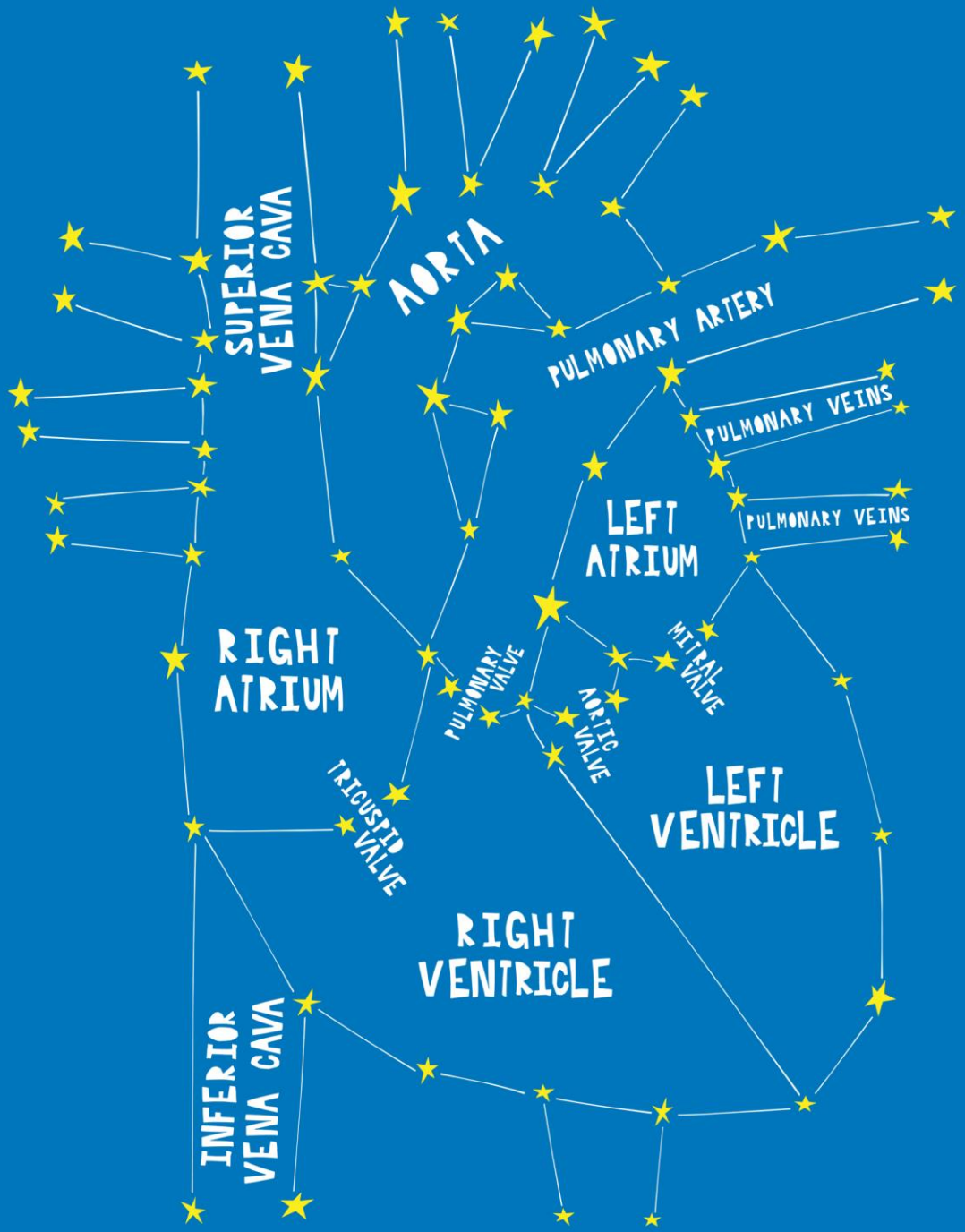
Prenatal Predictors of Outcomes in Congenital Heart Disease

David Saudek, MD

May 24, 2022



Disclosures

None

Objectives

- **For specific left heart and right heart lesions to understand:**
  - Pre- and post-natal physiology
  - Challenges in prenatal prediction of post-natal management
  - Short and long term outcomes

Topics

- Left heart
  - RV/LV size discrepancy
  - Hypoplastic left heart syndrome
- Right Heart
  - Tetralogy of Fallot

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Fetal Level of Concern

- LOC 1—Well baby nursery
  - Testing before discharge
- LOC 2--NICU
  - No PGE initially
  - Feed ad lib
- LOC 3—NICU
  - Start PGE at admission
  - High risk feeding protocol
- LOC 4--NICU
  - Cardiology at bedside
  - Cath team on standby(?)
- LOC 5—CICU
  - CICU attends delivery
  - Surgical team on standby

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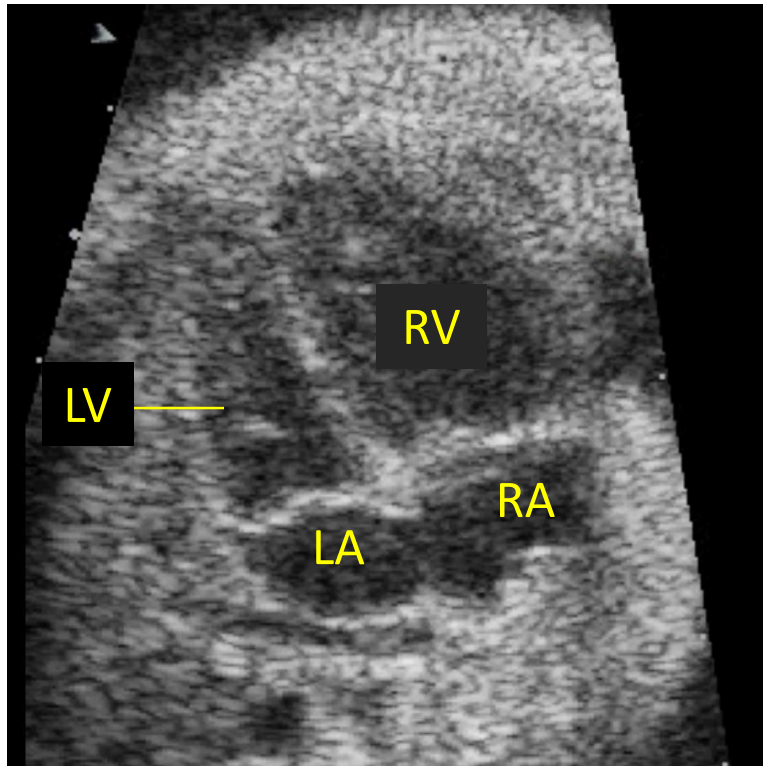
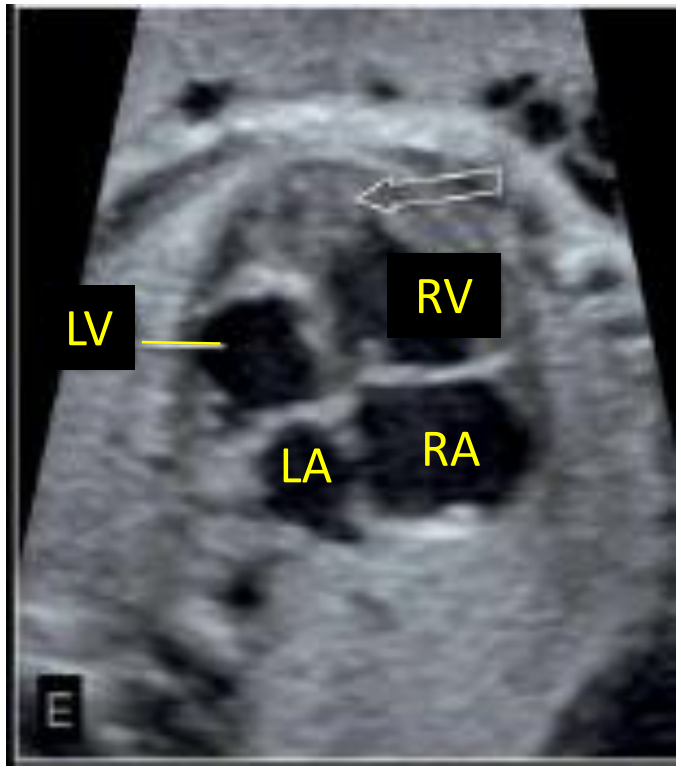
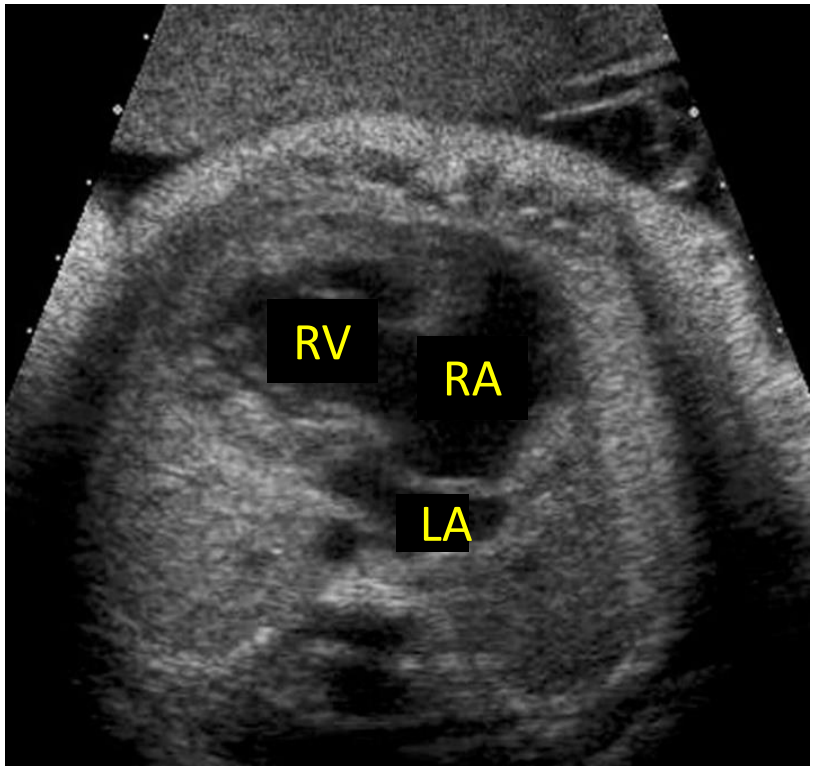
Spectrum of Left Heart Hypoplasia







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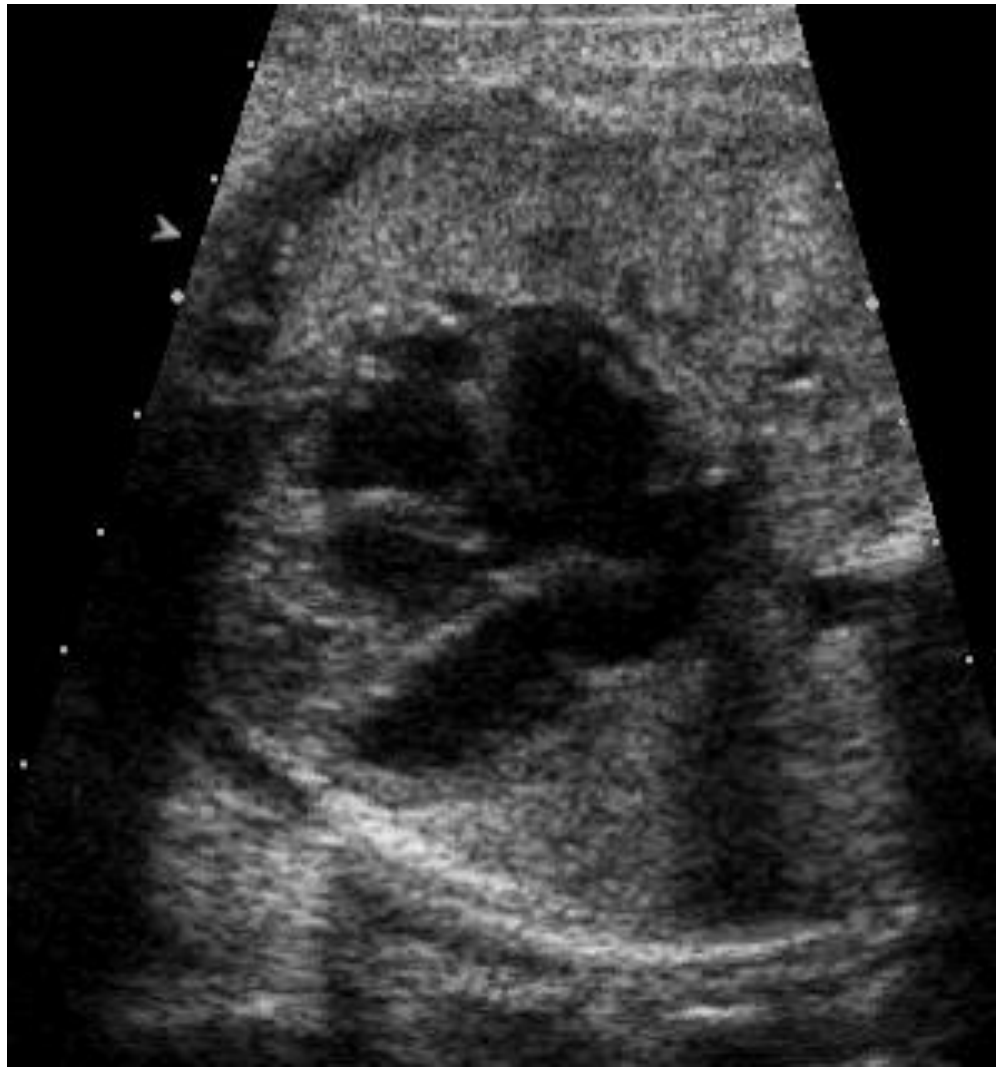
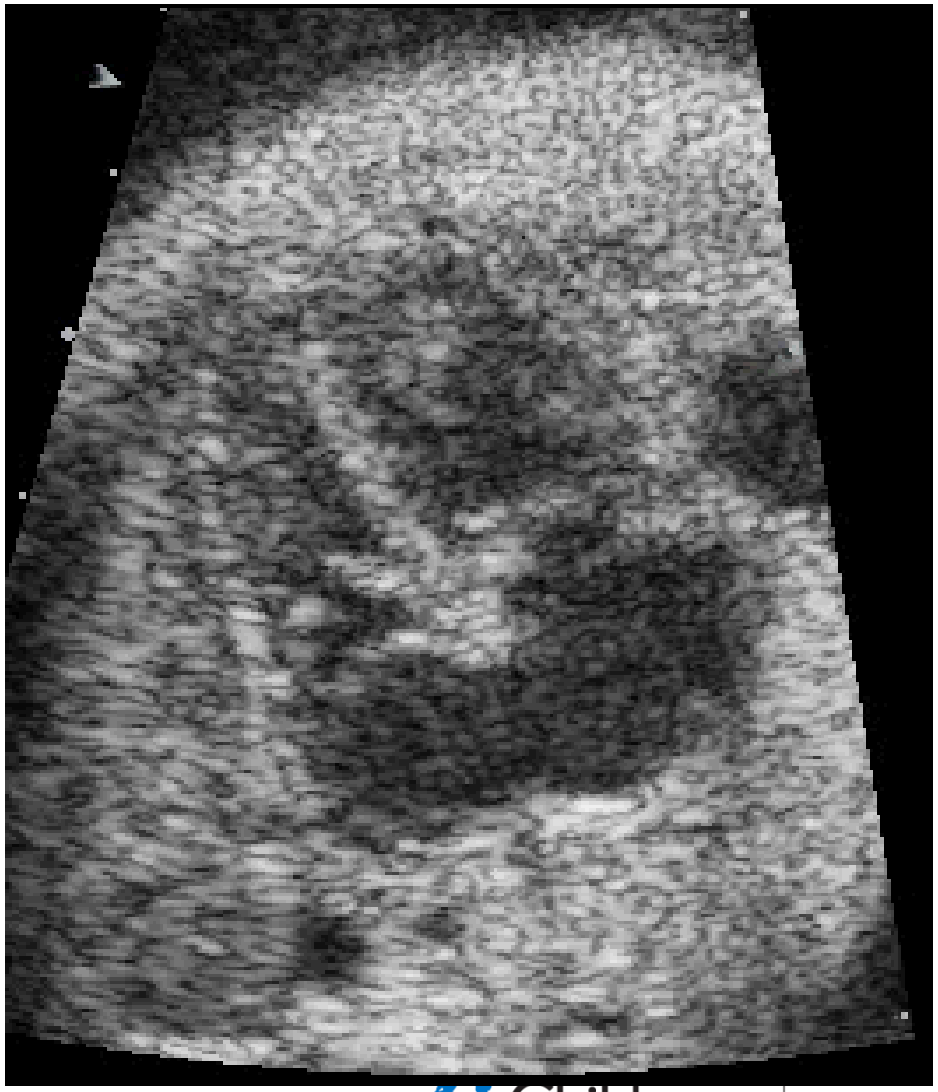
Baby with Coarctation



Baby with Normal Heart



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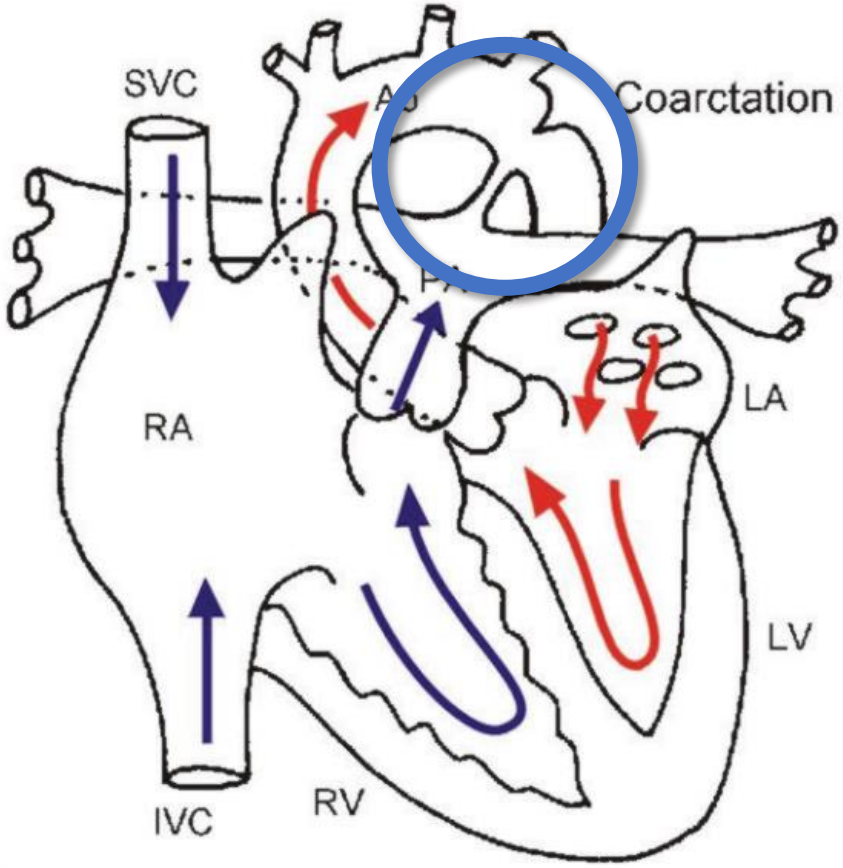

Isolated coarctation of the aorta in the fetus: A diagnostic challenge

Joshua A. Kailin MD | Alexia B. Santos MD | Betul Yilmaz Furtun MD | S. Kristen Sexson Tejtel MD, PhD, MPH | Regina Lantin-Hermoso MD

Toward Improving the Fetal Diagnosis of Coarctation of the Aorta

Meaghan Beattie[1] · Shabnam Peyvandi[1] · Suguna Ganesan[1] · Anita Moon-Grady[1]

Right Ventricular Enlargement In Utero: Is It Coarctation?

Shanthi Sivanandam[1] · Jessica Nyholm[1] · Andrew Wey[1] · John L. Bass[1]

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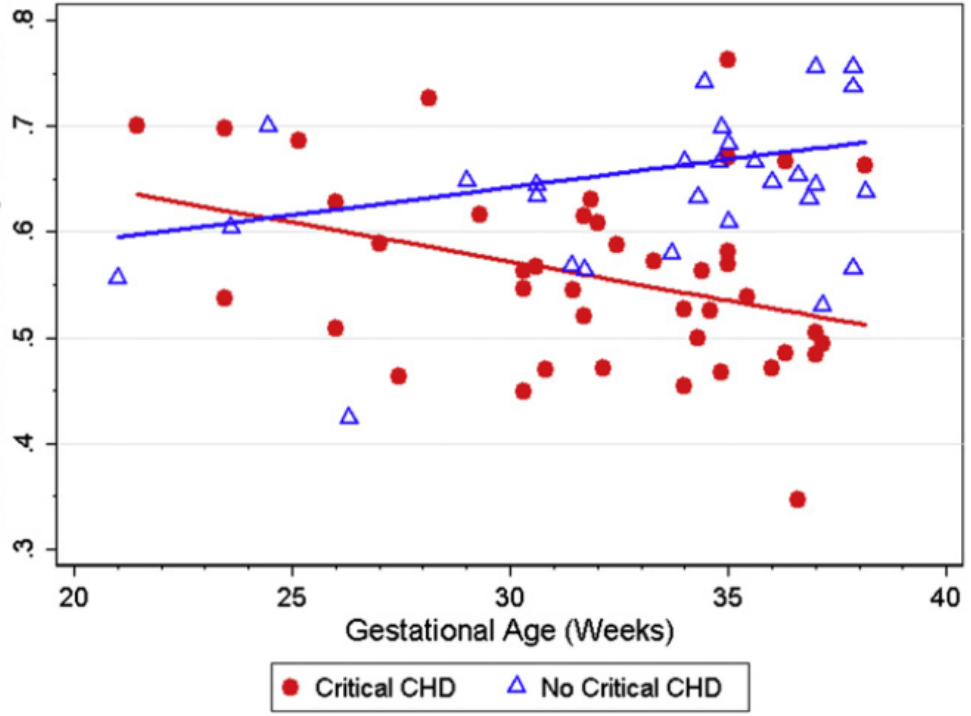
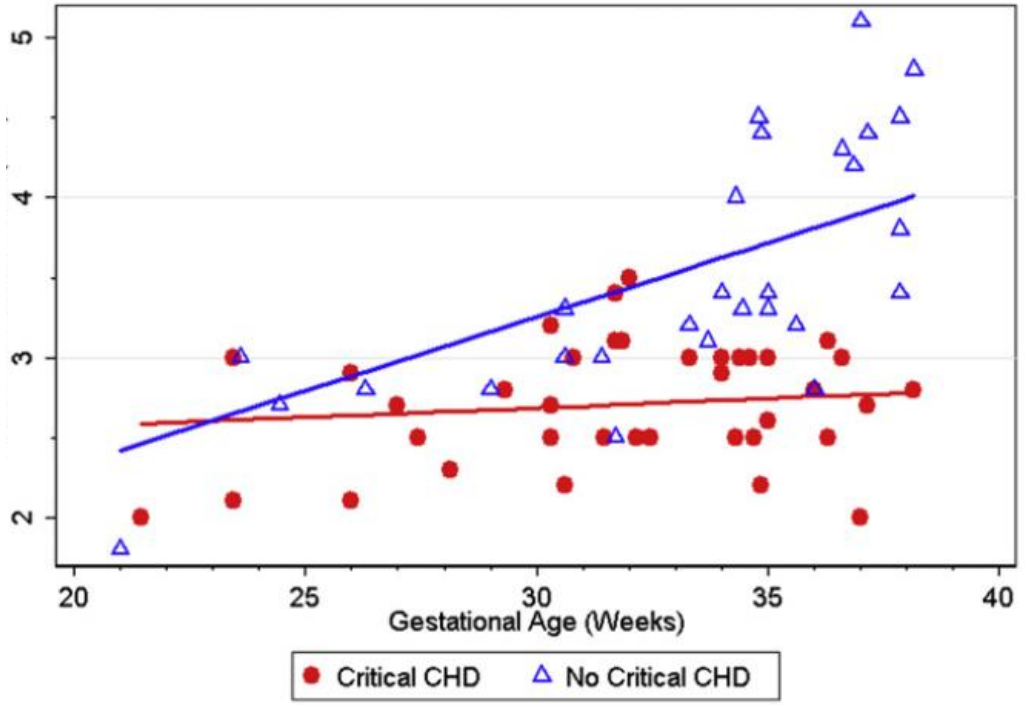
Ratio of Aortic valve: Pulmonary valve diameter



Aortic Arch Diameter



Parameter	Sensitivity	Specificity	PPV	NPV
MV/TV < 0.6	70%	87%	87%	68%
AV/PV < 0.6	79%	80%	83%	75%
LVmc/RVmc < 0.6	70%	67%	73%	62%
MV/TV or LVmc/RVmc < 0.6	95%	53%	73%	89%
[a]Arch ≤ 3 mm	94%	93%	83%	91%
[b]PFO BID or LTR	70%	100%	100%	68%

Quartermain, *Journal of the American Society of Echocardiography* 2009;22(11):1296-1301

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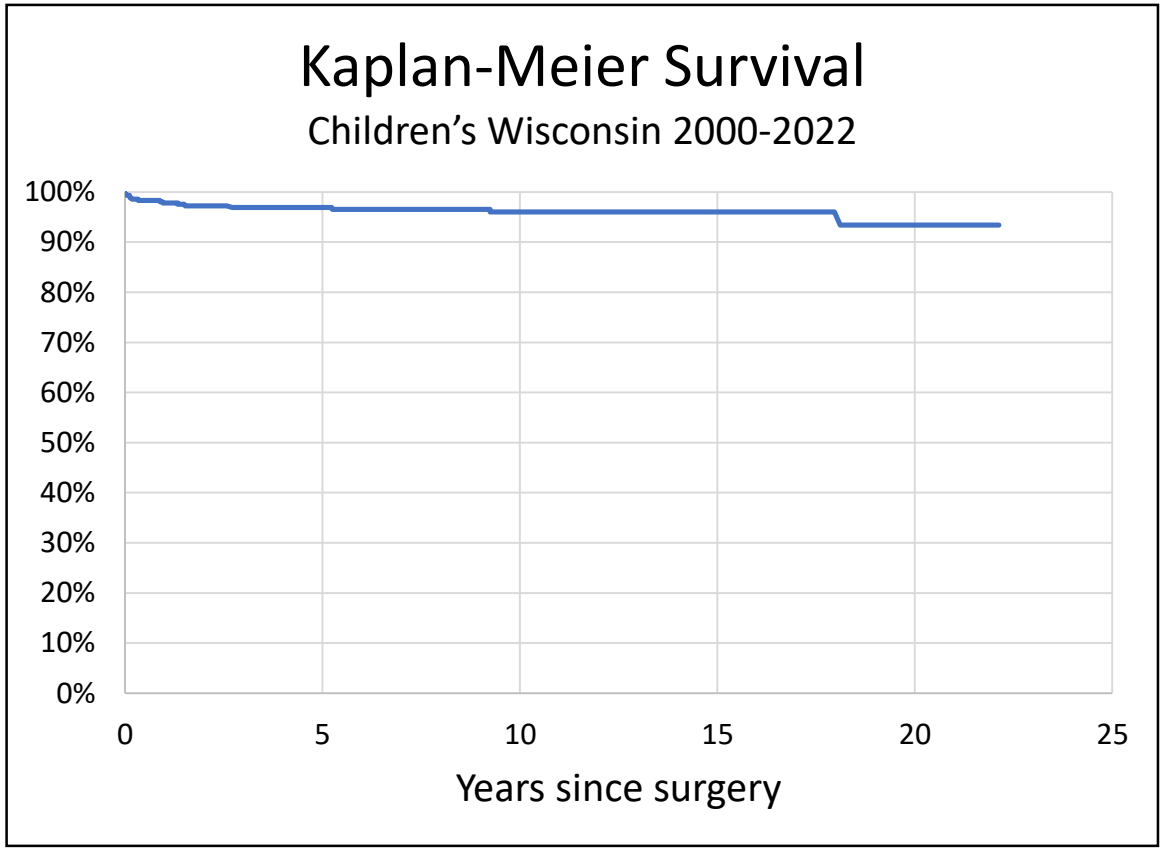
Coarctation Outcomes

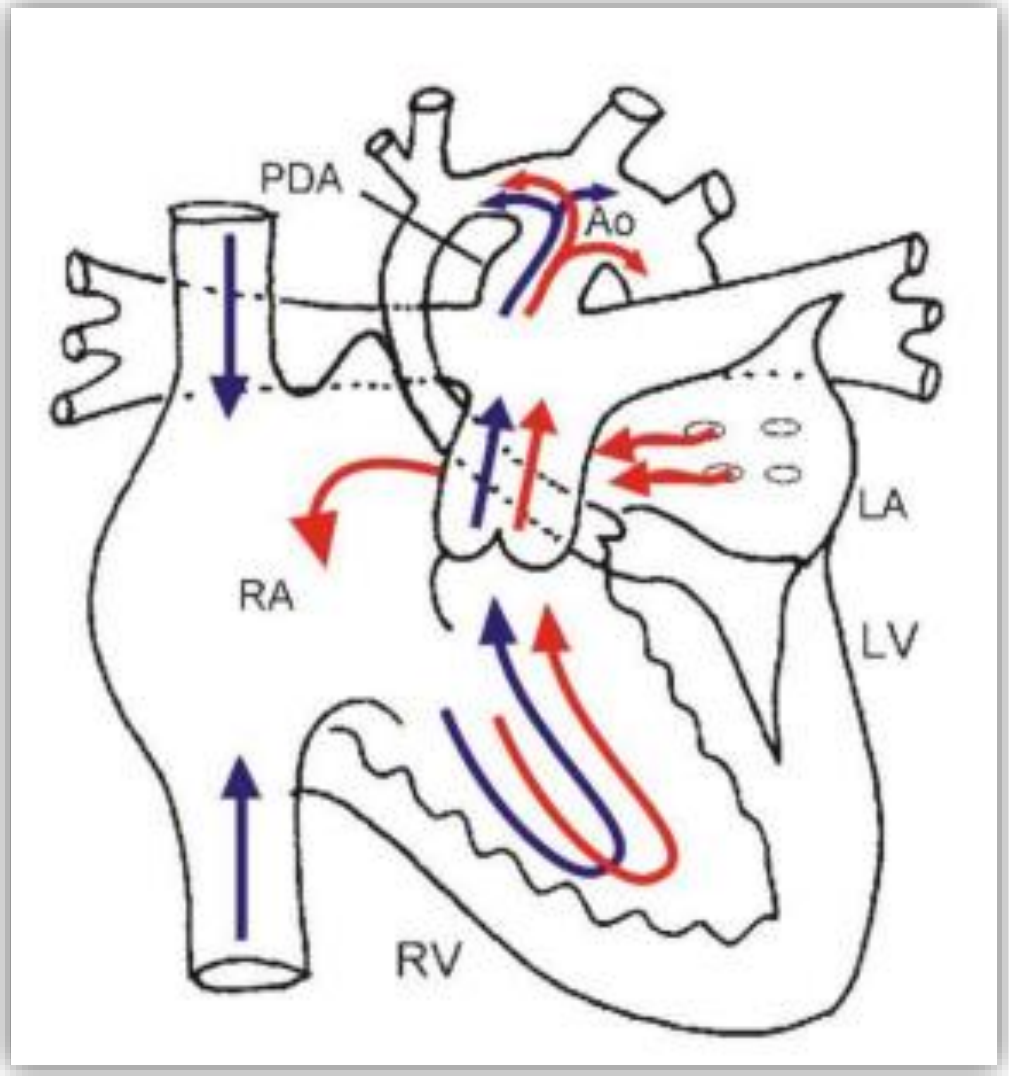
- Fetal LOC 1/2
  - Nursery vs NICU
  - No PGE
- Repair at time of diagnosis
- STS Operative Survival = 98.9%
- Median LOS = 7 days



Surgeries at Children's Wisconsin
n=456
93.4% survival at 22 years

Hypoplastic Left Heart

- Left ventricle inadequate to support systemic circulation
- “Flow equals grow”
- Requires single ventricle palliation
  - Norwood procedure—neonate
  - Glenn procedure—3-6 months
  - Fontan procedure—2-4 years



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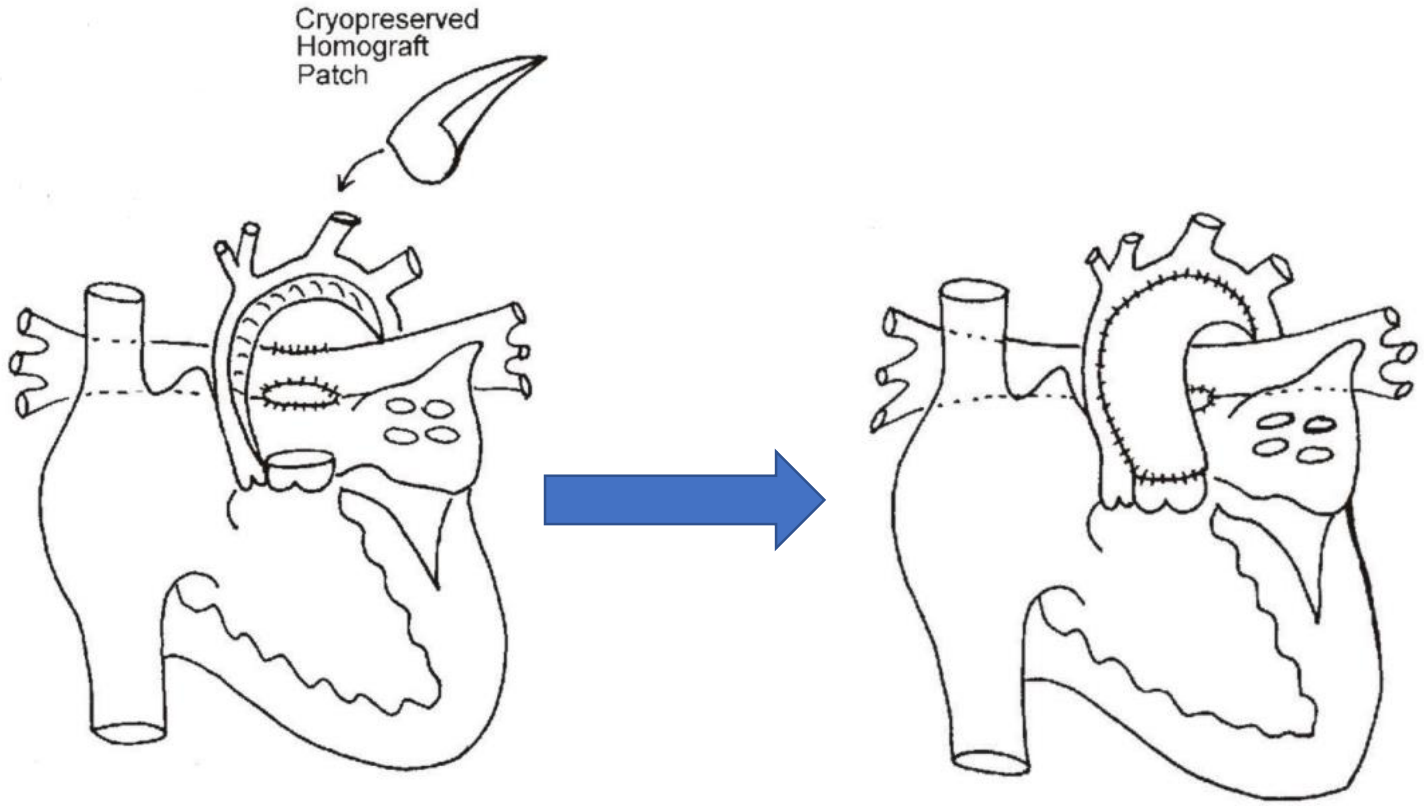
Norwood Procedure

- Goals:
  1. Unobstructed systemic blood flow
  2. Limited but adequate pulmonary blood flow
  3. Unrestricted pulmonary venous return



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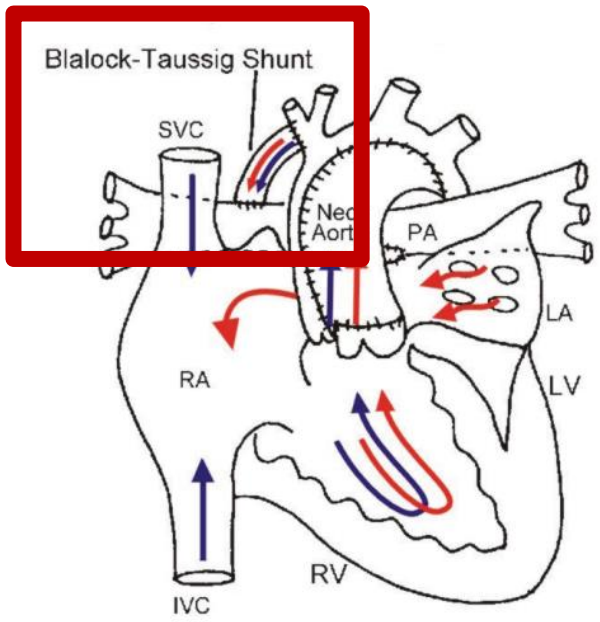
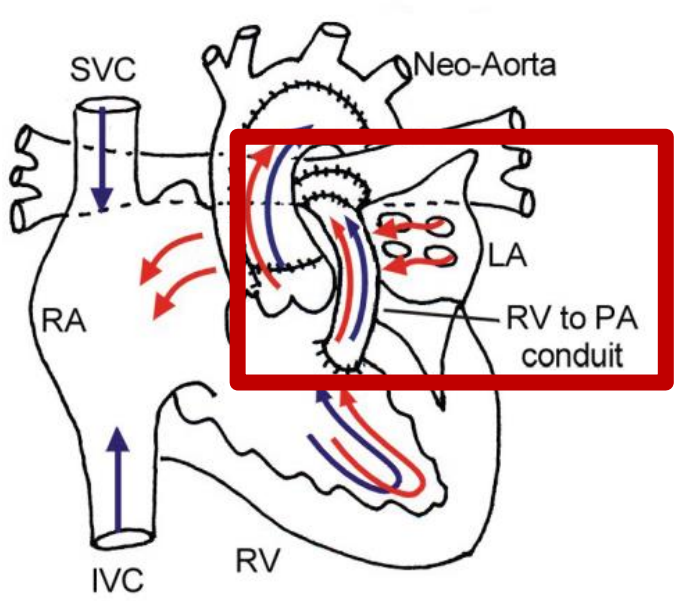
Norwood Procedure

- Goals:
  1. Unobstructed systemic blood flow
  2. Limited but adequate pulmonary blood flow
  3. Unrestricted pulmonary venous return

Either



or…



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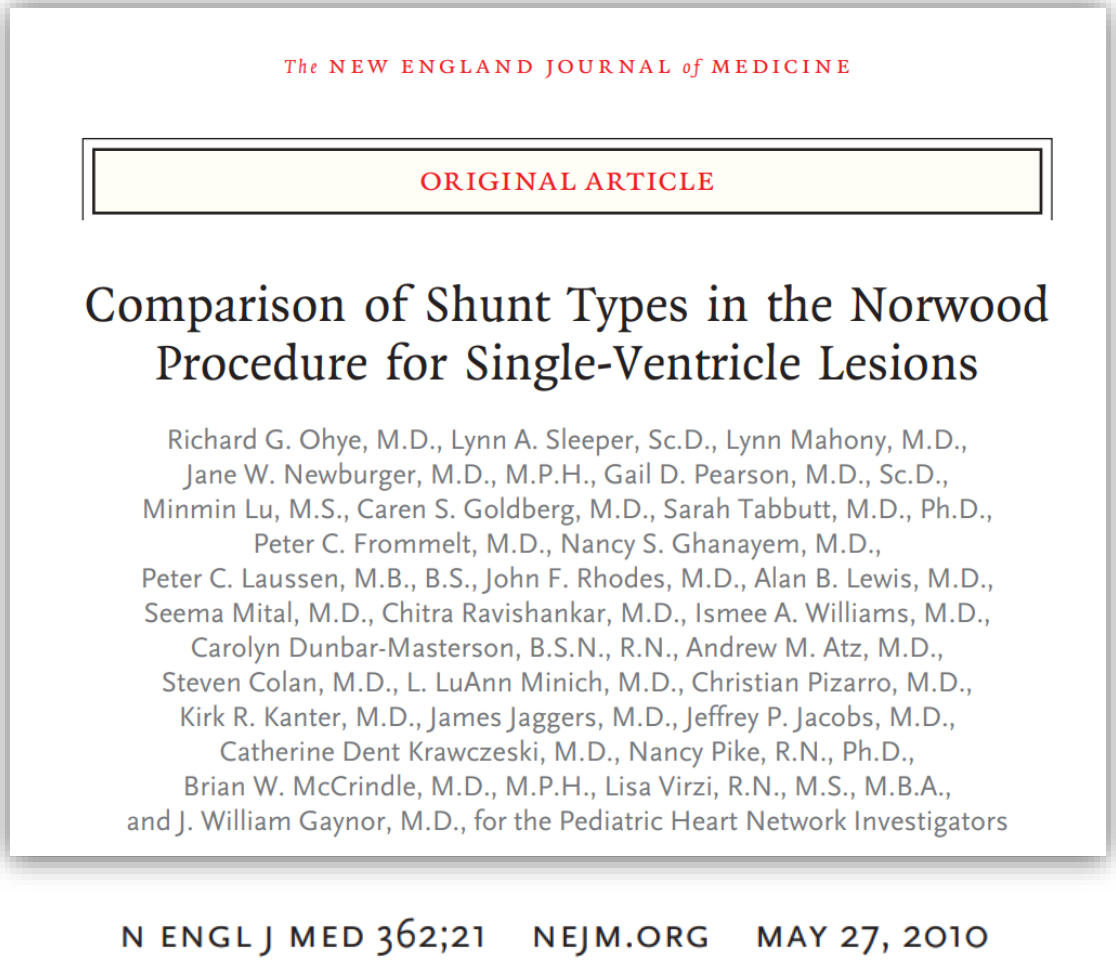

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Comparison of Shunt Types in the Norwood Procedure for Single-Ventricle Lesions

Richard G. Ohye, M.D., Lynn A. Sleeper, Sc.D., Lynn Mahony, M.D., Jane W. Newburger, M.D., M.P.H., Gail D. Pearson, M.D., Sc.D., Minmin Lu, M.S., Caren S. Goldberg, M.D., Sarah Tabbutt, M.D., Ph.D., Peter C. Frommelt, M.D., Nancy S. Ghanayem, M.D., Peter C. Laussen, M.B., B.S., John F. Rhodes, M.D., Alan B. Lewis, M.D., Seema Mital, M.D., Chitra Ravishankar, M.D., Ismee A. Williams, M.D., Carolyn Dunbar-Masterson, B.S.N., R.N., Andrew M. Atz, M.D., Steven Colan, M.D., L. LuAnn Minich, M.D., Christian Pizarro, M.D., Kirk R. Kanter, M.D., James Jaggers, M.D., Jeffrey P. Jacobs, M.D., Catherine Dent Krawczeski, M.D., Nancy Pike, R.N., Ph.D., Brian W. McCrindle, M.D., M.P.H., Lisa Virzi, R.N., M.S., M.B.A., and J. William Gaynor, M.D., for the Pediatric Heart Network Investigators

N ENGL J MED 362;21 NEJM.ORG MAY 27, 2010

- 549 infants undergoing Norwood
  - 15 pediatric heart centers
  - Randomized to RV-PA conduit or BTT shunt
  - RV-PA conduit had lower mortality at 12mo
    - 26.3% vs 36.4% mortality
  - Groups no different beyond 12 mo

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Prenatal diagnosis and risk factors for preoperative death in neonates with single right ventricle and systemic outflow obstruction: Screening data from the Pediatric Heart Network Single Ventricle Reconstruction Trial*

Andrew M. Atz, MD,[a] Thomas G. Travison, PhD,[b] Ismee A. Williams, MD, MS,[c] Gail D. Pearson, MD, ScD,[d] Peter C. Laussen, MBBS,[e] William T. Mahle, MD,[f] Amanda L. Cook, MD,[g] Joel A. Kirsh, MD,[h] Mark Sklansky, MD,[i] Svetlana Khaikin, RN, MPH,[h] Caren Goldberg, MD,[j] Michele Frommelt, MD,[l] Catherine Krawczeski, MD,[m] Michael D. Puchalski, MD,[n] Jeffrey P. Jacobs, MD,[o] Jeanne M. Baffa, MD,[p] Jack Rychik, MD,[q] and Richard G. Ohye, MD,[k] for the Pediatric Heart Network Investigators

The Journal of Thoracic and Cardiovascular Surgery • Volume 140, Number 6

Intermediate-term mortality and cardiac transplantation in infants with single-ventricle lesions: Risk factors and their interaction with shunt type

James S. Tweddell, MD,[a] Lynn A. Sleeper, ScD,[b] Richard G. Ohye, MD,[c] Ismee A. Williams, MD, MS,[d] Lynn Mahony, MD,[e] Christian Pizarro, MD,[f] Victoria L. Pemberton, RNC, MS,[g] Peter C. Frommelt, MD,[a] Scott M. Bradley, MD,[h] James F. Cnota, MD,[i] Jennifer Hirsch, MD, MS,[c] Paul M. Kirshbom, MD,[j] Jennifer S. Li, MD, MHS,[k] Nancy Pike, RN, PhD,[l] Michael Puchalski, MD,[m] Chitra Ravishankar, MD,[n] Jeffrey P. Jacobs, MD,[o] Peter C. Laussen, MBBS,[p] and Brian W. McCrindle, MD, MPH,[q] for the Pediatric Heart Network Investigators

The Journal of Thoracic and Cardiovascular Surgery • Volume 144, Number 1

- Risk for pre-operative mortality
  - Major extracardiac abnormality
  - Gestational age
  - Low birth weight
  - Obstructed pulmonary veins

- Risk for post-operative mortality
  - Genetic syndrome
  - Anatomic subtype (non-HLHS)
  - Lower Socioeconomic Status
  - Obstructed pulmonary veins

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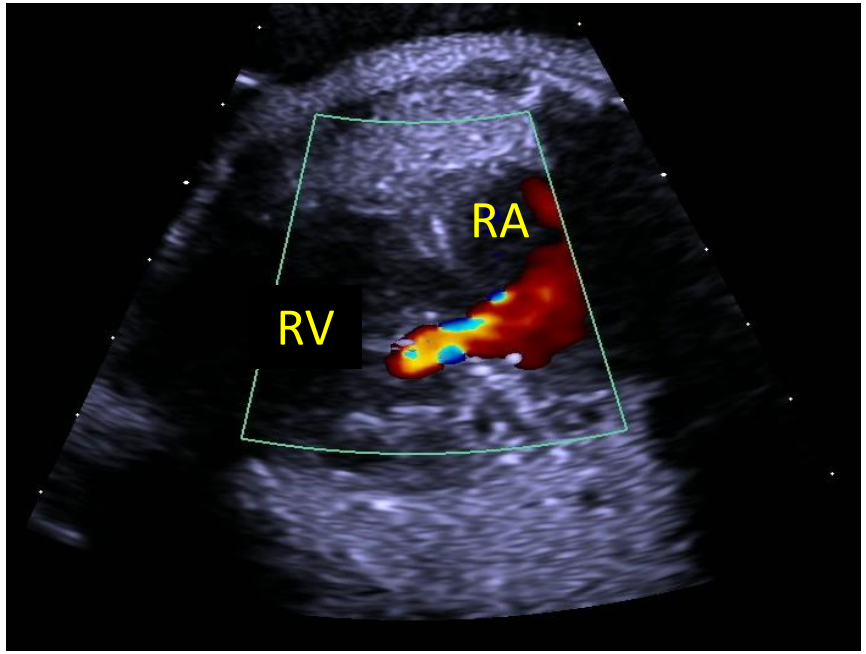
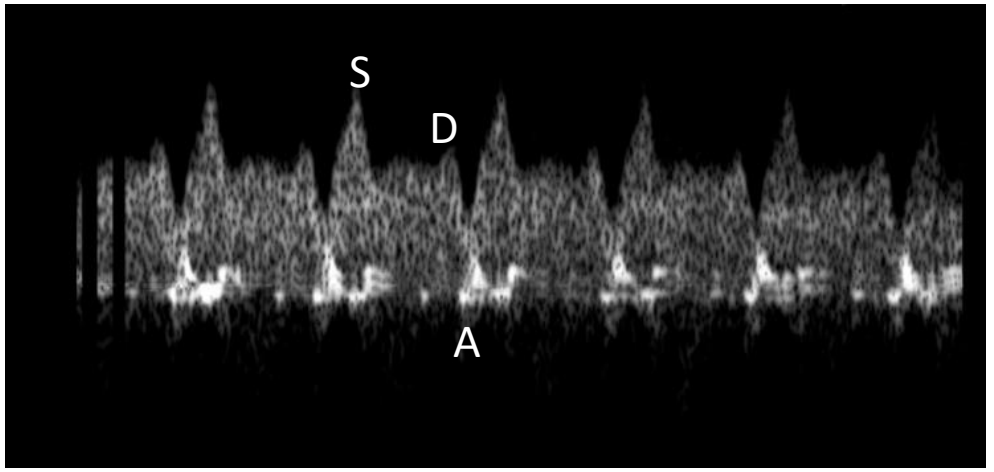
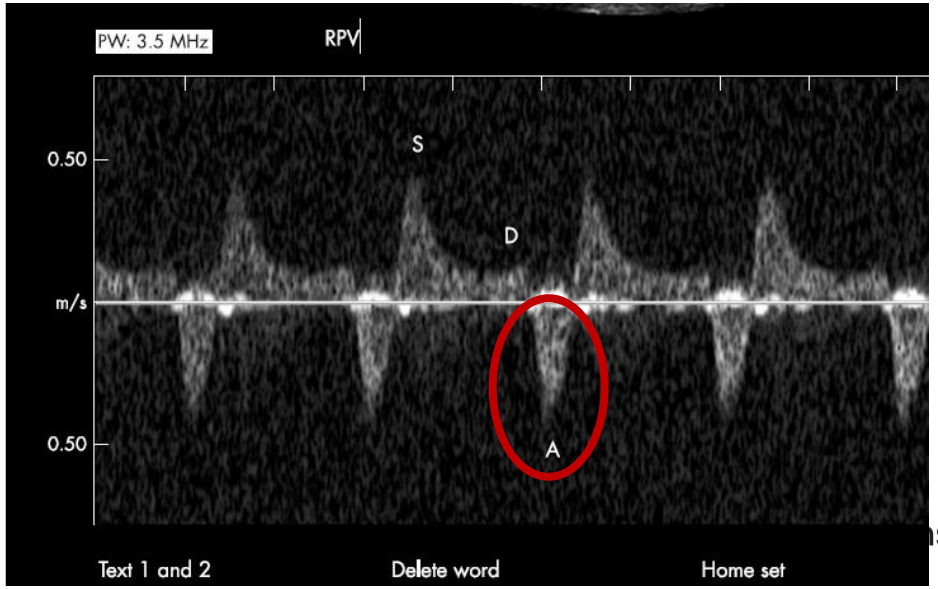
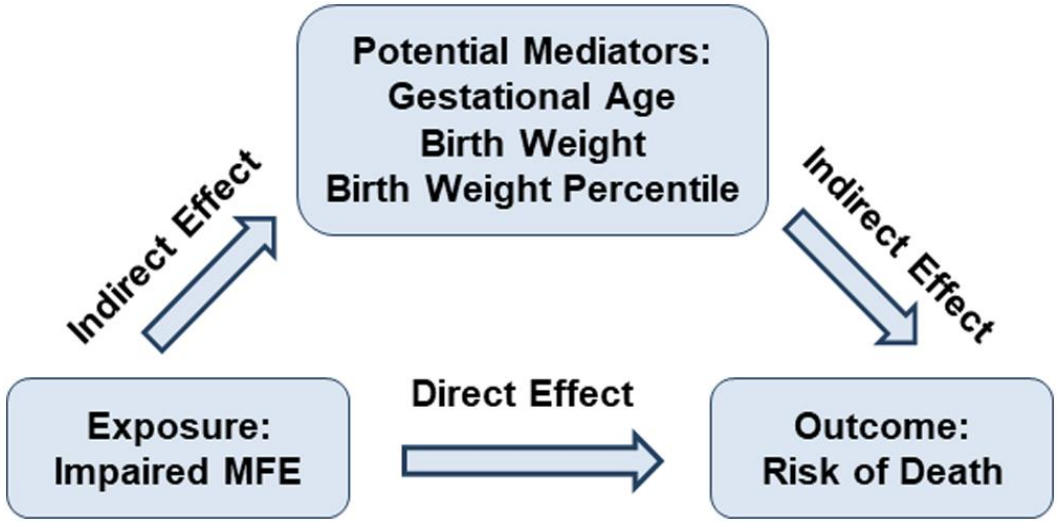




Mortality risk

- Restrictive atrial septum = 3.6x
- AV valve regurgitation ≥moderate = 3.6x





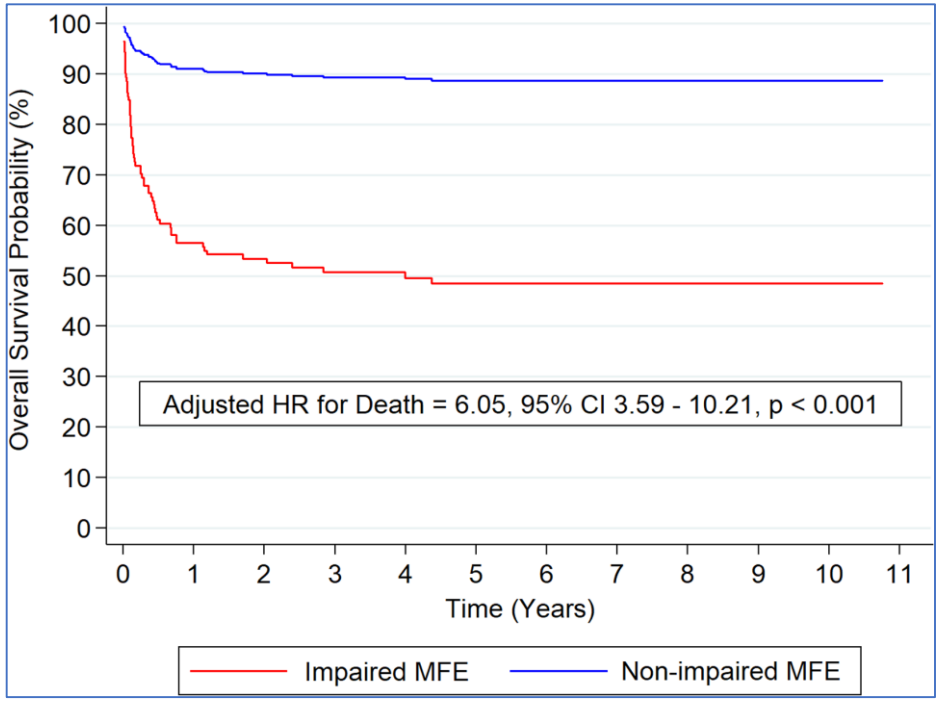


Table 2. Postoperative Details and Additional Surgeries

	Total cohort, N=273	Without impaired MFE, N=201	With impaired MFE, N=72	*P* value
Stage 1 Norwood procedure				
Survived to initial hospital discharge	235 (86)	187 (93)	48 (67)	<0.001
Length of postoperative hospital stay from surgery to discharge home, d	20 (14–37)	19 (13–32)	28 (17–46)	0.03

Jill J. Savla. Journal of the American Heart Association. Impact of Maternal–Fetal Environment on Mortality in Children With Single Ventricle Heart Disease, Volume: 11, Issue: 2

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Neonatal Approach to HLHS

- **Unobstructed pulmonary venous return**
  - Level of Concern 3
  - NICU for initial evaluation
  - Initiate PGE
  - Norwood at 5-7 days
- **Concerns for obstructed pulmonary venous return**
  - Level of Concern 5
  - NICU and CICU attend delivery
  - Surgical team on standby for emergent intervention

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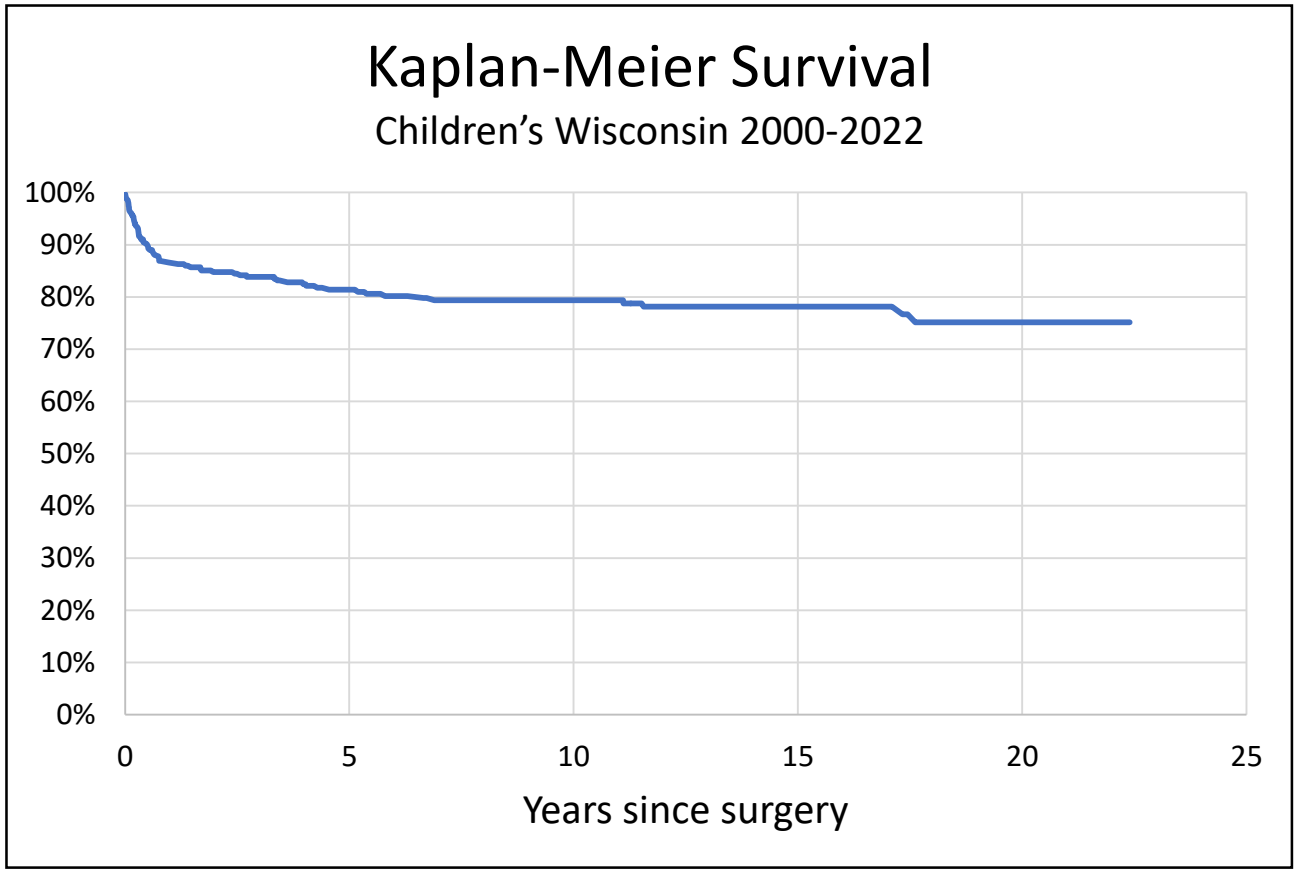
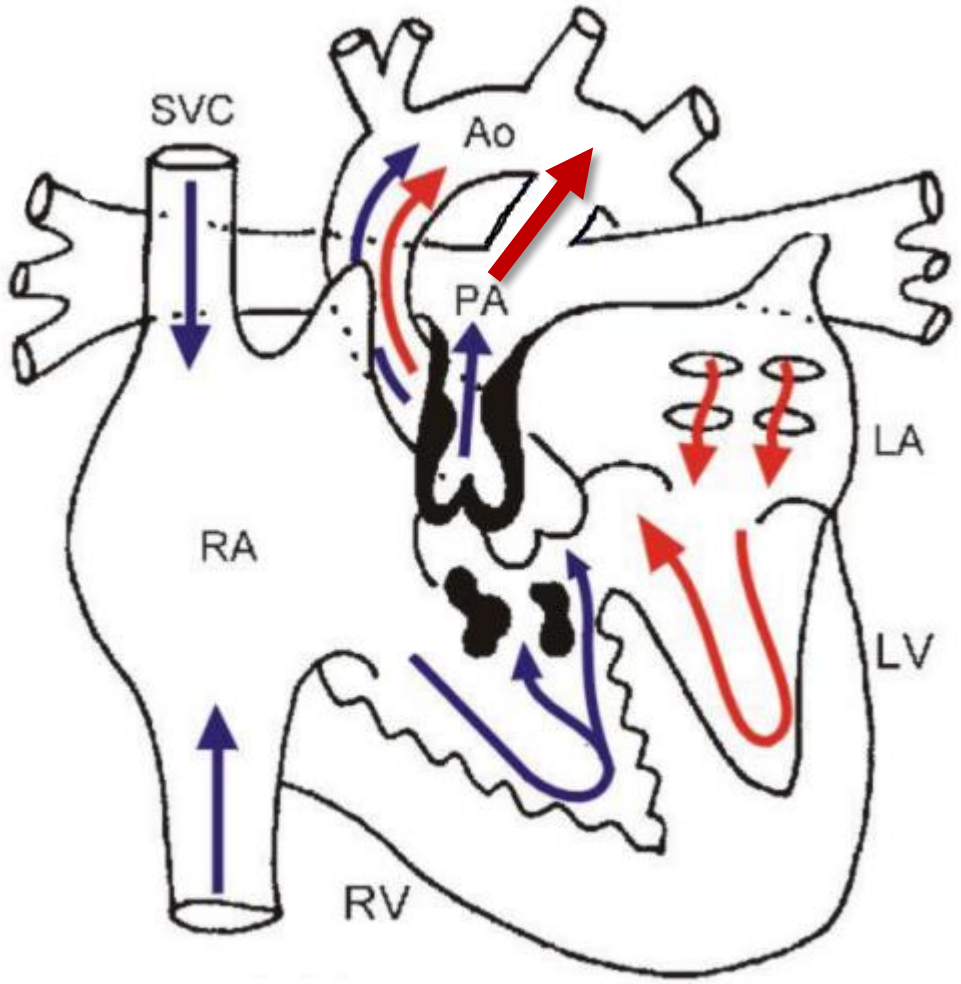
Norwood Outcomes

- STS Operative Survival = 88.2%
- Median LOS = 57 days
- CW 1 year transplant-free survival = 70%



Surgeries at Children's Wisconsin
n=334
75.2% survival at 20 years

Tetralogy of Fallot



Normally 8-10% of combined cardiac output goes to lungs

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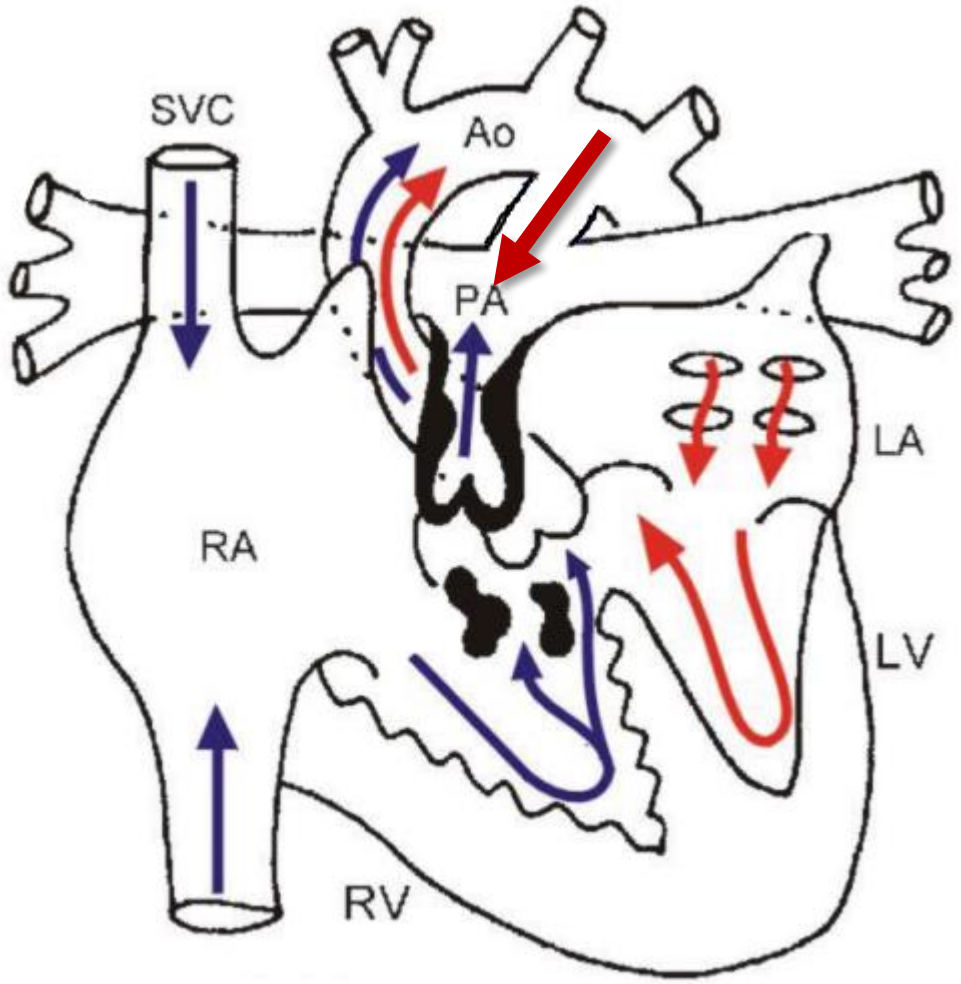
Tetralogy of Fallot



If inadequate pulmonary blood flow, ductal flow is left to right

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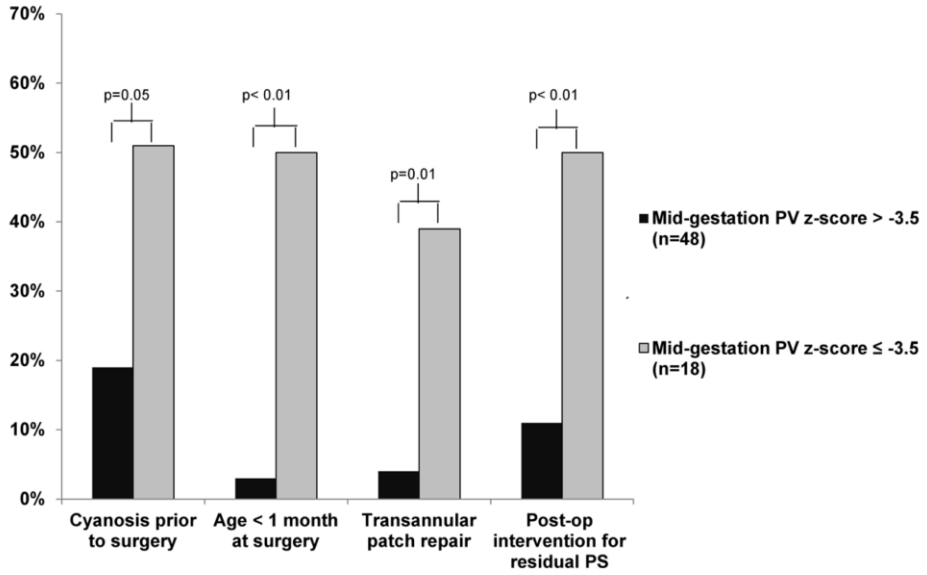
Predicting Neonatal Intervention



Parameter	*Sensitivity (%)*	*Specificity (%)*
PV-Z-score < −3 or PV/AoV ratio < 0.6		
Early-gestation (< 24 weeks)	100	50
Mid-gestation (24–32 weeks)	100	48
Final echo (mean age, 34 weeks)	92	50
Ductal flow pattern		
Early-gestation (< 24 weeks)	75	100
Mid-gestation (24–32 weeks)	88	94
Final echo (mean age, 34 weeks)	100	95

Concerning findings

- Pulmonary valve z-score < -3
- PV/AoV ratio <0.6
- PDA left to right in utero

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Neonatal approach to Tetralogy

- Likely adequate pulmonary blood flow
  - Fetal LOC 1/2
  - Watch PDA closure
  - Delay surgery until ~6mo
- Likely ductal dependent
  - Fetal LOC 2
  - Watch PDA closure
  - Neonatal intervention

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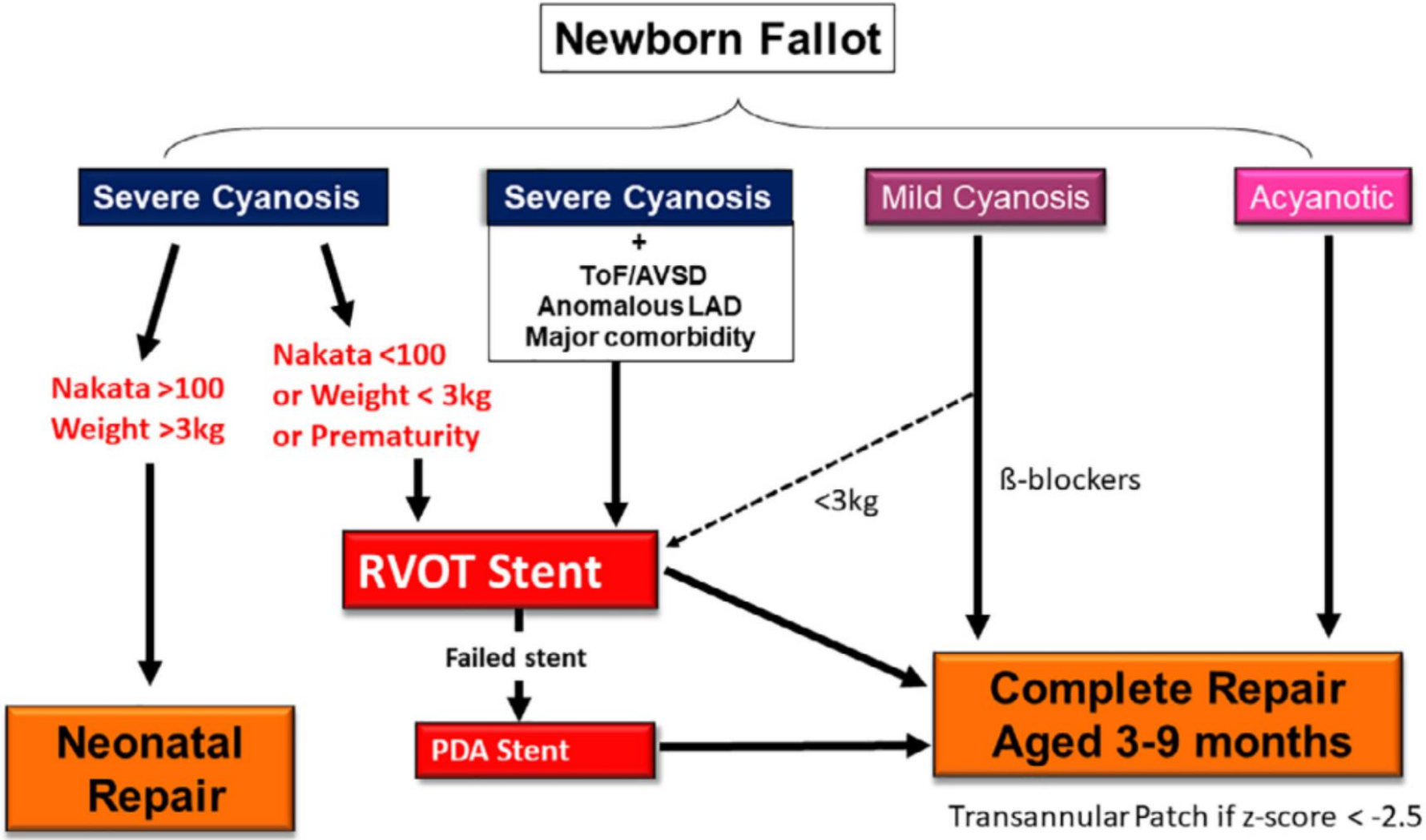

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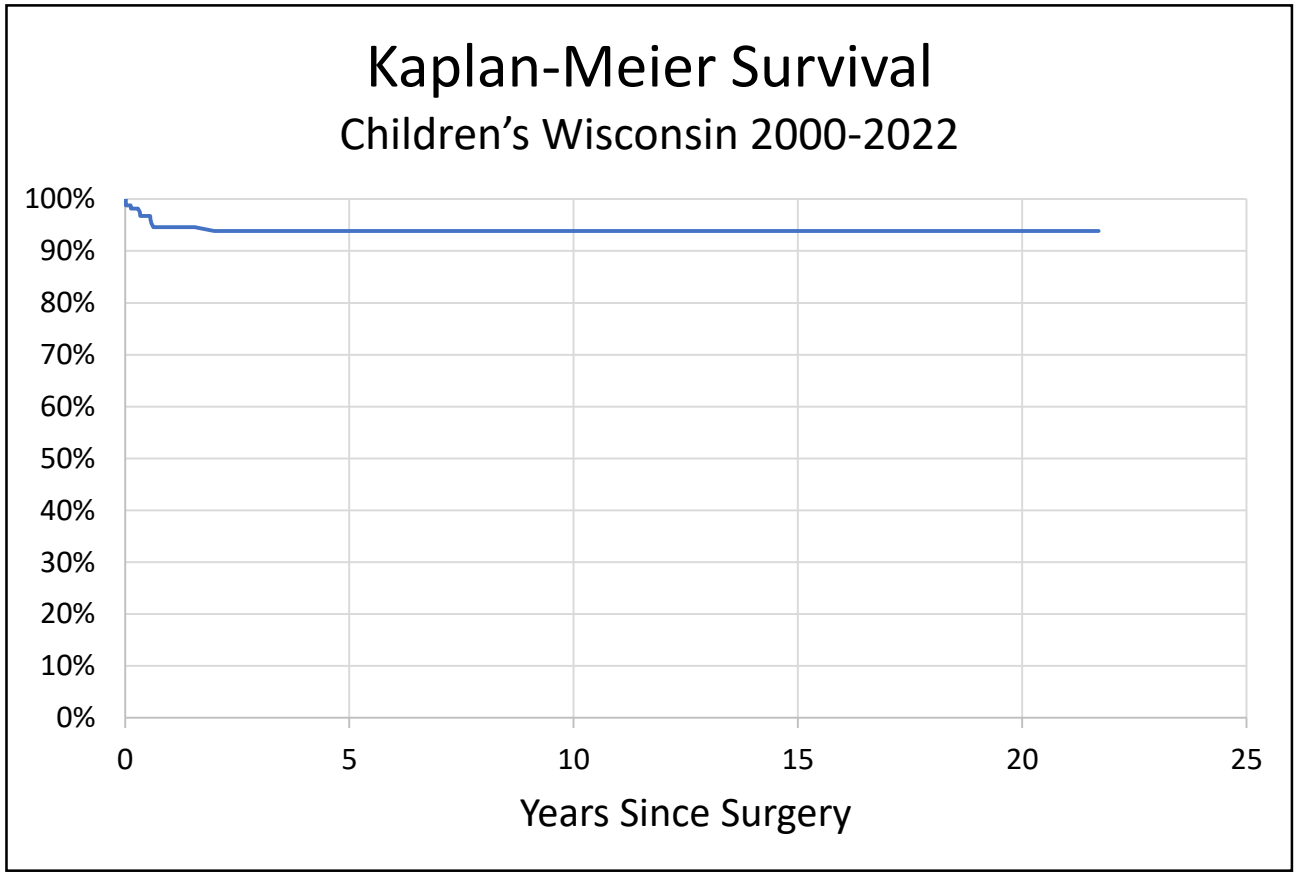
Tetralogy of Fallot Outcomes

- STS Operative Survival = 98.9%
- Median LOS = 12.6 days
- CW 20 year survival = 93.8%



Surgeries at Children's Wisconsin
n=160

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FETAL INNOVATIONS
Conversation with the Experts
Children's Wisconsin

Thank you!