Vanishing Twin Syndrome

Purpose and Goal: CNEP # 2091

- Understand vanishing twin syndrome in pregnancy.
- Learn about how reabsorbed twins effect surviving twins.

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Requirements for successful completion:

- Successfully complete the post-test
- Complete the evaluation form

Date
• November 2018 – November 2020

Learning Objectives

• Describe the pathophysiology of vanishing twin syndrome.
• Describe the neonatal effects of vanishing twins on surviving twins.
• Identify 2 approaches for the diagnosis and treatment of chimeric karyotypes.

Introduction

• Vanishing twin syndrome occurs:
  • With a set of twins
  • With a set of multiples
• It occurs when a fetus disappears in the uterus
• The timing of this event determines the outcome:
  • For the mother
  • For the surviving fetus

Vanishing Twin Syndrome

• Vanishing twin syndrome was first noted in 1945
• It is the identification of multi-fetal gestation:
  • With subsequent disappearance of a fetus
  • The fetus can be reabsorbed by:
    • The mother
    • The surviving fetus
• The overall rate of multi-fetal gestations:
  • Is 3-6% at 8 weeks gestation
• The rate of multi-fetal gestation:
  • Is higher at conception
• Than at the time of birth
• The loss if a twin can be expected:
  • In up to 7-40% of spontaneous pregnancies
  • In up to 27-38% of assisted pregnancies
• Vanishing twin syndrome:
  • Occurs in 21-30% of multi-fetal pregnancies
  • This holds true in the USA and Europe
• It has been described more frequently:
  • With early pregnancy ultrasounds
  • With in vitro fertilization (IVF)
    • IVF pregnancies are closely followed
    • The number of implanted eggs is known
• The timing of this event significantly:
  • Affects maternal complications
  • Affects the outcome of the viable twin

Pathophysiology

• The cause of a vanishing twin is unknown
  • Could be associated with congenital anomalies
  • Could be associated with abnormal implantation
• Abnormalities developing from a vanishing twin:
  • Usually appear early in development
  • As opposed to a later acute insult
• Placental or fetal analysis frequently reveals:
  • Chromosomal abnormalities
    • Diploidy
    • Triploidy
    • Abnormal sex chromosomes
• Chromosomal abnormalities may be found in:
  • Skin biopsies
  • Placental pathology
  • Chorionic villi sampling
• Chromosomal analysis of the surviving twin:
  • Is generally normal
• It is widely thought that the vanishing twin:
• Had a chromosomal abnormality
• That resulted in the disappearance
• With vanishing twin syndrome there may be:
  • Complete reabsorption of the fetus
  • Formation of a fetus papyraceus
    • A “mummified” fetus
    • A compressed fetus
  • Development of a subtle placenta abnormality
    • Placental cyst
    • Subchorionic fibrin
    • Amorphous material

Maternal Morbidity

• First trimester morbidity is limited
  • Mild vaginal bleeding
  • Mild uterine cramping
• Second and third trimesters
  • Premature labor
    • Significantly increases
    • Preterm <37 weeks
    • Very preterm <32 weeks
  • Infection from retained fetus
  • Severe puerperal hemorrhage
  • Consumptive coagulopathy
  • Obstruction of labor
    • Low lying papyraceus
    • Causing labor dystocia
    • Leading to Cesarean section
  • Emotional stress from loss of twin
    • Grief and bereavement

Fetal Morbidity and Mortality

• The surviving fetus
- Loses a twin
- Is at risk for hypotension
  - At the time of disappearance
- Is at risk for poor perfusion
  - At the time of disappearance
- Is at risk for skin necrosis
  - Cutis aplasia
  - Results from hypotension
- Is at risk for cerebral palsy
- Complications are generally low
  - If the event occurs early
- Complications can be more severe
  - If the event occurs in the first trimester
    - The surviving twin can be flattened
    - From vanishing amniotic fluid absorption
    - From vanishing placental tissue absorption
  - If the event occurs in the third trimester
    - Miscarriage of both twins can occur

**Neonatal Complications**

- Prematurity
  - There is a high risk of prematurity
  - At least 23% are born premature
  - Many are born <32 weeks gestation
- Low birth weight or SGA
  - There is a high risk of low birthweight
  - At least 33% are born SGA
  - Many require NICU care
- Chimeric genetic disorders
- Cerebral palsy (CP)
  - There is an increased risk of CP
  - Related to release of thromboplastic proteins
    - Absorption of proteins by surviving twin
    - Which leads to reverse blood flow
    - Which leads to coagulopathy
• Which leads to CNS damage
• Related to hypotension from reverse blood flow
  • Which leads to IVH
• Congenital anomalies
  • Microcephaly
  • Hydrocephaly
  • Eye anomalies
  • Cleft lip/palate
  • Cardiac anomalies

Chimeric Genetic Disorders

• A chimeric genetic disorder refers to:
  • Possession of more than one genetic identity
  • It can refer to animals or humans
• The term chimera comes from ancient mythology
  • Chimeras were incredible beasts
  • They were a composite of two creatures
    • With a lion’s head
    • And a serpent’s tail
• In humans, chimeras are composites of two embryos
  • The embryos becomes fused in utero
  • This is considered to be very rare
• Chimeric disorders are known as 46 XX/46 XY karyotypes
• They are caused by having two distinct cell populations
• They arise from the combination of an XX zygote and an XY zygote
• The two zygotes would have normally developed into twins
  • 46 XX/46 XY is associated with ambiguous genitalia
• Physical symptoms vary widely from infant to infant
• The most common symptoms are:
  • A small phallus
    • Midway in size
    • Between a clitoris and a penis
  • An incompletely closed urogenital opening
    • Or a shallow vagina
  • An abnormal urethra opening
• Located on the perineum
• Infants possess both ovarian and testicular tissue
  • The ovaries may function fully, partly or not at all
  • The testes may function fully, partly, or not at all
• Segmentation of skin (distinct patches) may be seen
• Different colored eyes may also be seen (this is rare)
• 46 XX/46 XY does not involve cognitive impairment
• At puberty, male and female characteristics may emerge
• Genetic testing is the only reliable method of diagnosis

Differences in Sexual Development

• Infants with ambiguous genitalia
  • Should be considered gender neutral
  • Should be referred for specialty care
• The Differences in Sexual Development Clinic
  • Urology
  • Endocrinology
  • Genetics
  • Cytogenetics
  • Gynecology
  • Psychiatry
  • Adolescent Medicine
• Diagnosis is based on several studies:
  • Physical examination
  • DNA studies
  • Cytogenetic studies
  • Hormonal studies
  • Radiographical evaluation
  • Ultrasonography evaluation
  • Endoscopic studies
  • Laparoscopic studies
  • Psychosocial assessment
• Treatment involves several approaches:
  • Hormonal treatment
  • Nonsurgical treatment
• Vaginal dilation
• Surgical treatment
  • Creation of genitals
  • Genital reconstruction
• Psychosocial support
  • Long term follow up and support
• Goals of treatment include:
  • Education
    • Family
    • Healthcare providers
• Counseling and support
  • A conflict of identity should be anticipated
  • Sexual confusion should be anticipated
  • Sexual disorientation should be anticipated

Summary

• Vanishing twin syndrome is relatively common
• There is little *physical* effect on the mother
• There can be considerable effect on the surviving twin
  • Human chimeric disorders may be seen
• All infants with ambiguous genitalia
  • Should be considered gender neutral
  • Should be referred for specialty care
  • DNA studies should guide sex assignment

References


Evaluation

Required fields

Your information

*Your name

*Your email address

*Your Seattle Children's ID

*Your hospital
  - Seattle Children's Hospital
  - Providence Regional Medical Center Everett
  - Overlake Medical Center
  - St. Joseph Medical Center
  - St. Francis Hospital
  - Harrison Medical Center
  - Other Medical Center or Hospital

Test

- Vanishing twin syndrome was first identified in 1945.
  - True
  - False

- Vanishing twin syndrome is the reabsorption of a twin in utero.
  - True
  - False

- Vanishing twin syndrome occurs in up to 30% of pregnancies.
• More twin gestations are present at conception than at delivery.
  ○ True
  ○ False

• Surviving twins are commonly SGA and may have congenital anomalies.
  ○ True
  ○ False

• Ambiguous genitalia may arise from a chimeric karyotype in which both male and female DNA are present.
  ○ True
  ○ False

**Evaluation**

We hope you found this educational offering both interesting and informative. We’d like to hear from you and appreciate you taking the time to answer these evaluation questions.

*Were you able to complete this activity in the allotted time?*
  ○ Yes
  ○ No

Were you informed of the following disclosures?

*Purpose of learning activity*
  ○ Yes
  ○ No
*Requirements for successful completion of this CNE activity
- Yes
- No

*Presence or absence of conflict of interest of planning committee members
- Yes
- No

*Presence or absence of conflict of interest of content specialist(s)/author(s)/feedback person(s)
- Yes
- No

*Were your personal objectives successfully achieved?
- Yes
- Somewhat
- No
If not, why not?

*What one thing might you do differently in your practice after this session?

Please evaluate the brief:

*Presentation organized
- Excellent
- Very good
- Good
- Fair
- Poor
*Materials offered relevant content
  ○ Excellent
  ○ Very good
  ○ Good
  ○ Fair
  ○ Poor

*Assistance provided as needed
  ○ Excellent
  ○ Very good
  ○ Good
  ○ Fair
  ○ Poor
  ○ Not applicable

*Overall strength of presentation
  ○ Excellent
  ○ Very good
  ○ Good
  ○ Fair
  ○ Poor

Stated objectives achieved?

*Describe the pathophysiology of vanishing twin syndrome.
  ○ Excellent
  ○ Very good
  ○ Good
  ○ Fair
  ○ Poor

*Describe the neonatal effects of vanishing twin syndrome on surviving twins.
*Identify 2 approaches for the diagnosis and treatment of chimeric karyotypes.

- Excellent
- Very good
- Good
- Fair
- Poor