

Neonatal Nursing Education Brief: Vanishing Twin Syndrome

<https://www.seattlechildrens.org/healthcare-professionals/education/continuing-medical-nursing-education/neonatal-nursing-education-briefs/>

Vanishing twin syndrome is becoming more common with the increased availability of in vitro fertilization. More twins exist at the time of conception than at the time of delivery. Surviving twins are at risk for being SGA as well as for developing both male and female characteristics.

NICU, IVF pregnancy, twin gestation, surviving twin, congenital anomalies

Vanishing Twin Syndrome

Purpose and Goal: CNEP # 2091

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

None of the planners, faculty or content specialists has any conflict of interest or will be presenting any off-label product use. This presentation has no commercial support or sponsorship, nor is it co-sponsored.

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
 - Diplody
 - 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

Anderson-Berry, A.L. 2016. Vanishing Twin Syndrome. Medscape: <http://emedicine.medscape.com/article/271818-overview>

Landy, H.J & Keith, L.G. 1998. The Vanishing Twin: A Review. Human Reproduction Update, 4 (2), p. 177-183.

Niu, D.M., Pan, C.C. Lin, C.Y., Hwang, B.T., and Chung, M.Y. 2002. Mosaic or Chimera? Revisiting an Old Hypothesis About the Cause of 46,XX/46,XY Hermaphrodite. Journal of Pediatrics, 140 (6), p. 732–735.

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.

- True
- False
- 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.

- Excellent
- Very good
- Good
- Fair
- Poor

*Identify 2 approaches for the diagnosis and treatment of chimeric karyotypes.

- Excellent
- Very good
- Good
- Fair
- Poor