




Ultrasound Differentiation of Twins with Discordant Congenital Diaphragmatic Hernia in the Delivery Room

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Abstract

Most infants with prenatally diagnosed congenital diaphragmatic hernia (CDH) are intubated rapidly after birth to optimize oxygenation and ventilation while avoiding abdominal distention and high mean airway pressures. A twin pregnancy complicated by one twin with a CDH diagnosis is a rare event and is associated with preterm delivery and low birth weight compared to singletons with CDH. In rare cases of discordant CDH in twin pregnancies with an absence of external distinguishing features (similar weights, fetal presentation, and sex), it may be difficult to quickly determine which twin has CDH in the delivery room (DR), raising ambiguity about the best management of both infants. This case describes the successful use of ultrasound (US) in the DR to rapidly diagnose the presence or absence of CDH in discordant twins. By developing a resuscitation algorithm and using in situ simulations prior to delivery, the twin with CDH was rapidly identified, intubated, and transported to the neonatal intensive care unit (NICU) for further management. The twin without CDH received routine care and was transferred to the well-baby nursery. Interprofessional planning and simulation may be used to design a safe resuscitation plan incorporating US diagnosis of diaphragmatic anomalies into the Neonatal Resuscitation Program (NRP) algorithm.

Keywords

- ▶ congenital diaphragmatic hernia
- ▶ ultrasound
- ▶ simulation
- ▶ delivery room
- ▶ twin gestation

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Introduction

Congenital diaphragmatic hernia (CDH) is a rare birth defect that allows abdominal viscera to herniate into the thoracic cavity. This defect can disrupt the physiological transition after birth, leading to hemodynamic instability and pulmonary hypertension.¹ European and Canadian groups have guidelines on delivery room (DR) management of CDH, based largely on expert opinion.²⁻⁴ The Neonatal Resuscitation Program (NRP) does not provide specific recommendations on DR management of infants with CDH.⁵ Typically, most are intubated immediately after birth to provide adequate ventilation and oxygenation while avoiding high airway pressures to reduce lung injury.⁶ Immediate intubation and targeted ventilation strategies, along with other maneuvers, are critical to reduce the risk of developing a life-threatening pulmonary hypertensive crisis and avoiding the need for extracorporeal membrane oxygenation (ECMO) prior to surgical repair.

CDH occurring in twin pregnancies is an even rarer event.⁷⁻¹⁰ Within these, there exists the possibility of either CDH concordance (both twins have CDH) or discordance (one twin with CDH and one twin without CDH). In discordant twins, external distinguishing markers may be prenatally known in the affected twin (sex, weight, other congenital malformations, prenatal birth order and presentation), and allow for confident differentiation between the babies after birth.^{11,12} However, the absence of such markers creates a potential conflict in DR management, specifically, whether immediate intubation of both twins should be performed to optimize care of the twin with CDH, at the expense of imposing an unnecessary intubation on the twin without CDH.

Prenatal birth order has been shown to be highly concordant with neonatal birth order (91.7%).¹² Performance of an ultrasound (US) just prior to delivery could improve this. However, there remain concerns regarding the accuracy of the maternal side and position following delivery through a small hysterotomy, particularly given the potential for serious harm from delayed intervention for the twin with CDH. A postnatal chest radiograph is another excellent method of diagnosing CDH, but there may be limitations to obtaining one fast enough in the DR to make real-time management decisions. Rapid identification of the presence or absence of CDH in the DR utilizing US may be another method of directly differentiating between CDH discordant twins and providing individualized care. This practice has not been previously described and the length of time required to make the diagnosis is unclear.

Case Presentation

A 30-year-old nulliparous woman spontaneously conceived dichorionic-diamniotic twins. Fetal anatomic survey performed at a gestational age (GA) of 20 weeks revealed normal anatomy in twin A (maternal right) and an isolated left-sided diaphragmatic hernia in twin B (maternal left). Both twins were noted to be female and growth assessments were appropriate for GA. Fetal magnetic resonance imaging (MRI) confirmed the findings of CDH in twin B, and the liver was suspected to be uninvolved. Follow-up US assessments

were notable for third trimester fetal growth restriction in both twins, with elevated umbilical artery (UA) Dopplers in twin B. Estimated lung-to-head ratio (LHR) was 1.68, associated with better prognosis in neonates with CDH.¹³ While twin A was persistently cephalic, twin B had variable presentations on serial assessment. Given the presence of CDH, elevated UA Dopplers in twin B, and growth restriction of both twins, the decision was made for an early term delivery via elective cesarean section (CS). Due to the variable presentation of twin B, in utero position was not a reliable feature to identify the affected twin.¹¹

An interprofessional team including obstetrics, maternal-fetal medicine, neonatology, and pediatric radiology was assembled to attempt US determination of CDH immediately after delivery. Previous literature on complex resuscitation scenarios guided the development of a resuscitation algorithm.^{14,15} Table-top simulations were utilized to identify the needs of each team, potential barriers, and safety concerns, and integrate these into a proposed DR resuscitation algorithm that incorporated radiology into the care of the newborns, but prioritized immediate patient needs over awaiting radiologic diagnosis (►Fig. 1). In situ simulation drills were conducted to review and iteratively refine the algorithm, determine the positioning of team members and equipment, and rehearse responses to clinical scenarios (►Fig. 2). On delivery day, “just in time,” in situ simulations were run to rehearse management and formalize communication phrases between team members.

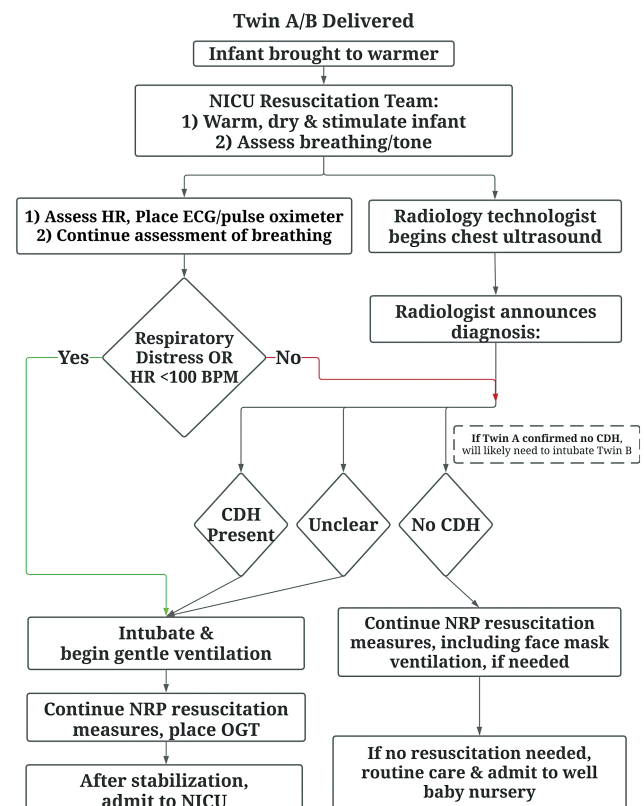


Fig. 1 Delivery room resuscitation algorithm. NICU, Neonatal Intensive Care Unit; HR, Heart Rate; ECG, Electrocardiogram; BPM, Beats Per Minute; CDH, Congenital Diaphragmatic Hernia; NRP, Neonatal Resuscitation Program; OGT, Orogastric Tube.

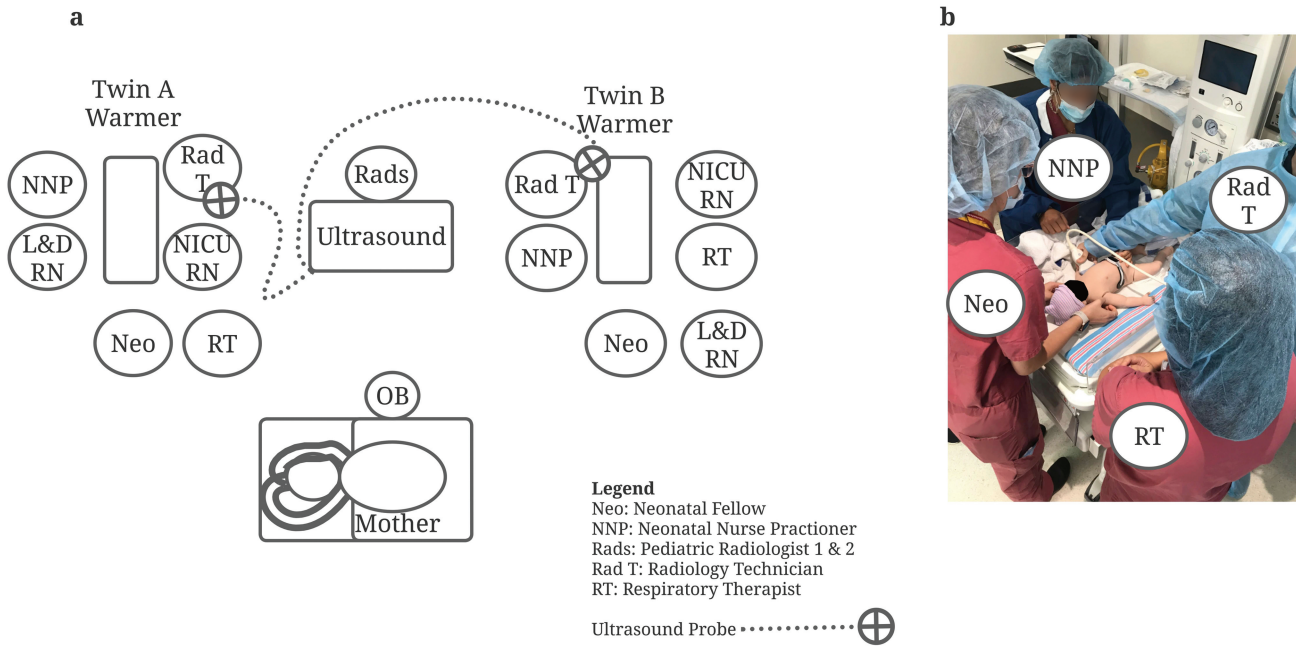


Fig. 2 (a) Graphic depiction of delivery room personnel and equipment positioning. (b) Photograph of in situ delivery room simulation with labeled clinical personnel. Legend included for abbreviations of personnel and equipment.

An elective CS was performed at 37 weeks of GA. A General Electric (GE) LOGIQ E10 machine with a high-resolution linear 6- to 15-MHz probe was used for US analysis. A protocol of two cine sweeps was developed. The chest was first scanned with the probe transverse midline at the level of the heart, sweeping inferiorly to the level of the diaphragm, with a goal to show the presence or absence of intra-abdominal organs adjacent to the heart and mediastinal shift. A second cine was obtained with the probe sagittal at the level of the midclavicular line, including the chest and top of the diaphragm in the field of view, and sweeping laterally. This view was intended to confirm the intact diaphragm with the stomach visualized below the level of the diaphragm or the presence of abdominal organs in the chest. Scanning was performed by a pediatric radiology technician (one assigned to each baby) and interpreted in real time by two pediatric radiologists. Absence of CDH was announced in twin A 45 seconds after delivery, with US diagnosis made within 19 seconds (→Fig. 3). The infant received routine postnatal care, was transferred to the well nursery, and discharged with the mother on the day of life (DOL) 4. Twin B was diagnosed with CDH at 47 seconds of life (US diagnosis made within 14 seconds), intubated by 68 seconds of life, and was stabilized and transferred to the NICU. Twin B underwent operative repair on DOL 3, was extubated on DOL 7, and weaned off respiratory support by DOL 13.

Discussion

To our knowledge, this is the first case describing the use of US in the DR management of discordant CDH in twins without external distinguishing features. It is important to develop strategies to manage twin infants with CDH discordancy as they are more likely to be born prematurely and

have lower birth weights than singleton pregnancies with CDH, leading to poorer overall outcomes and survival.¹⁰ Interestingly, CDH discordance in twins is more likely to occur than CDH concordance.⁸⁻¹⁰ There are several proposed mechanisms for this presentation, including epigenetic abnormalities, environmental factors, and genetic causes like incomplete penetrance or new dominant mutations.^{8,10}

Rapid intubation of the twin with CDH was the primary goal of our resuscitation. Like previously published reports of complex, patient-specific neonatal resuscitations, interprofessional predelivery planning, and in situ simulation drills led to the development of a resuscitation algorithm that prioritized the cardiopulmonary requirements of each infant.^{14,15} Of additional benefit, in situ simulation helped the team identify human factors needed to facilitate efficient and effective care, specifically the need to define and practice the use of various communication phrases during the delivery to create a shared mental model among the different disciplines while moving through the resuscitation algorithm. As a result, rapid and accurate US determination of the presence of CDH was achieved in 14 seconds and the absence of CDH in 19 seconds, optimally addressing the resuscitation needs of the twin with CDH and sparing the twin without CDH unnecessary intubation and NICU admission.

One potential limitation for general applicability is the relatively “mild” prenatal CDH presentation in this infant (favorable LHR and apparent lack of liver involvement). In more severe CDH presentations, 14 seconds to determine the presence of CDH may be considered too long to spend on US confirmation. Nonemergent deliveries of similar cases without external distinguishing features can be incorporated in utero predelivery US assessment to aid in increasing the predictive value of which twin will be delivered first. In these cases, detailed discussions regarding reliability must be

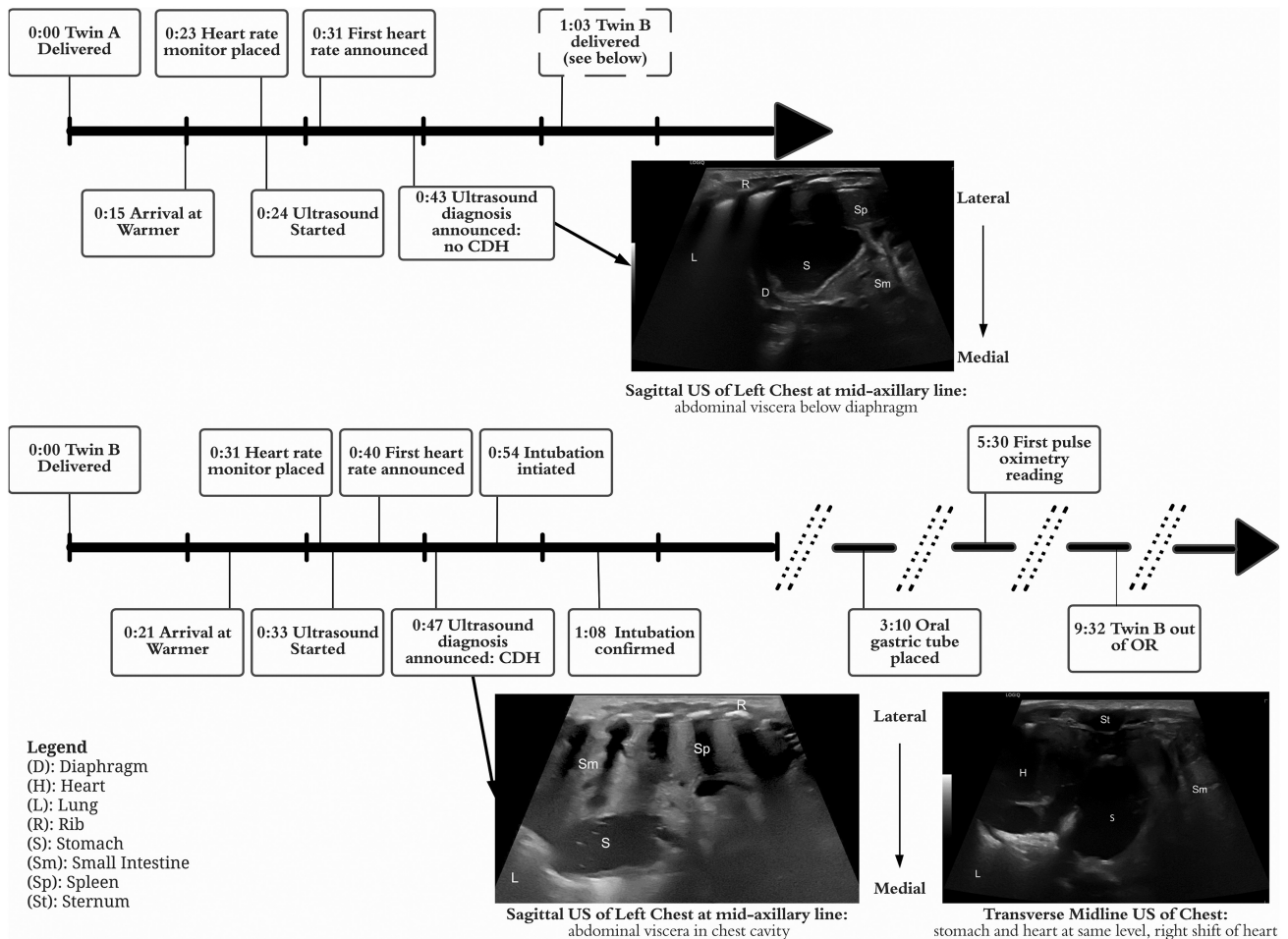


Fig. 3 Delivery room timeline of both twins with representative ultrasound images taken during resuscitation highlighting the presence or absence of congenital diaphragmatic hernia (CDH). Legend included abbreviations used in ultrasound labeling.

weighed if postnatal management will be contingent entirely on this assessment. Rapid postnatal US assessment of each twin remains the most accurate way to definitively assess the presence or absence of CDH, particularly given the need for immediate intervention and the serious consequences of delayed action.

This case illustrates the successful use of US to differentiate between twins with CDH discordancy in DR. With prior interprofessional preparation and simulation, effective use of US in high-risk delivery scenarios is possible.

Author Contributions

S.M.C wrote the first draft of the manuscript with specific contributions from B.R. and S.H.S.M.C, E.K., C.P., C.C., S.C., A.K., B.R., and C.O.-H. contributed to the conception and design of the study, critical revision of the manuscript for important intellectual input, gave final approval of the manuscript, and agree to be accountable for all aspects of the work.

Study Approval Statement

This study protocol was reviewed and approved by Weill Cornell Medicine Institutional Review Board (approval number: 21-12024204).

Patients' Consent

The parents of these children provided written informed consent for publication of this case report and use of the images therein.

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None.

Conflict of Interest

None declared.

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