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Pre-intervention mortality in ectopia cordis: an underrecognised phase of care

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Letter to the Editor

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To the editor,

Ectopia cordis is a rare congenital anomaly in which the heart is partially or completely displaced outside the chest cavity, with an estimated incidence of 5–8 per million live births.[1] Despite advances in prenatal diagnosis, neonatal resuscitation, and surgical procedure of CHDs, ectopia cordis carries substantial mortality, particularly in the pre-intervention phase.[2] Infants who die before operative intervention remain underrepresented, resulting in an incomplete understanding of the true disease burden.

From 2023 to 2026, three neonates with ectopia cordis were referred to our tertiary congenital cardiac centre. None had a prenatal diagnosis, and all were delivered outside specialised facilities. During the study period, fetal anomaly screening and referral pathways for complex congenital cardiac anomalies were still evolving in our setting. Limited access to specialised fetal echocardiography, variable availability of antenatal anomaly scans, and delayed referral likely contributed to the absence of prenatal diagnosis.

Each infant was born either term or near term, and their respective birth weights ranged from 1.8 to 2.5 kg. One had thoracic ectopia cordis, while the remaining two had thoracoabdominal variants. Early mortality appeared multifactorial, driven by severe haemodynamic compromise at presentation, delayed referral, absence of immediate sterile protective coverage of exposed cardiac structures, hypothermia, and infection risk. In each case, the window of opportunity for successful surgical intervention was missed prior to being able to achieve the level of physiologic stability required to safety perform surgery.

Reported survival in ectopia cordis remains poor despite advances in care. Large series have demonstrated overall survival rates of approximately 10%, with neonatal mortality approaching 50–60% even in well-resourced settings with prenatal diagnosis and planned delivery.[4] Importantly, these reported outcomes primarily reflect patients who survive long enough to undergo surgical evaluation or intervention. Our experience highlights an even earlier and often unreported phase of mortality, suggesting that the true burden of disease may be substantially underestimated.

Contemporary management of ectopia cordis emphasises staged approaches involving early protective coverage, gradual reduction of mediastinal contents, and delayed intracardiac repair following physiological stabilisation. However, these advances benefit only infants who survive the immediate postnatal period and are delivered in centres equipped for coordinated perinatal and surgical care. In resource-limited or diagnostically delayed settings, the absence of prenatal identification often results in unplanned delivery and suboptimal early management.

Our experience suggests that pre-intervention mortality represents a critical yet underrecognised phase in the care of infants with ectopia cordis. Addressing this gap requires strengthening prenatal detection, improving referral networks, and implementing standardised neonatal stabilisation protocols, which may substantially increase the number of infants who survive to undergo definitive surgical reconstruction. Furthermore, multicentre registries capturing both preoperative mortality and surgical outcomes are essential for accurately defining disease burden, informing parental counselling, and guiding future improvements in care.

Although limited by small sample size, reporting fatal experiences alongside surgical successes provides a realistic depiction of ectopia cordis management and underscores the importance of early, coordinated perinatal and neonatal care in this challenging condition.[2,3] Improving the pre-intervention phase may ultimately determine which infants reach the point where surgical reconstruction becomes feasible.

Sincerely,
Dr. Kajal Khatri
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