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Initial Stabilization Strategies for a Newborn with Larsen Syndrome and Meconium Aspiration Syndrome in a Peripheral Hospital

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Abstract
Larsen syndrome is a rare congenital skeletal dysplasia characterized by multiple joint dislocations and distinctive craniofacial features, which can complicate neonatal airway management. We report a term female neonate born at 40 weeks of gestation, weighing 2200 grams, delivered spontaneously in a peripheral facility with thick meconium-stained amniotic fluid. The infant did not cry at birth and exhibited severe respiratory depression (Apgar scores 3–4–5). Initial resuscitation, including airway clearance and positive pressure ventilation without a self-inflating bag, was performed to achieve effective lung aeration, resulting in gradual improvement in oxygen saturation from 93% to 100%. Clinical examination revealed congenital knee dislocation and cephalhematoma, while anthropometric assessment indicated intrauterine growth restriction. Chest auscultation demonstrated no adventitious sounds despite persistent tachypnea. This case highlights the complexity of neonatal stabilization when meconium aspiration syndrome coexists with syndromic skeletal anomalies. In resource-limited settings, structured resuscitation protocols and careful handling of the airway and limbs are essential to optimize outcomes and prevent further injury.

Keywords*: Larsen syndrome, Congenital joint dislocation, Neonatal meconium aspiration*

INTRODUCTION

Globally, neonatal mortality remains a critical public health challenge, with approximately 2.3 million deaths occurring in the first 28 days of life annually, accounting for nearly half of all under-five mortality worldwide (Amuka et al., 2020; Guerrera, 2015; Hug et al., 2019; Moradi-Lakeh et al., 2018; Tessema et al., 2023). Among the leading causes of neonatal death are birth asphyxia, respiratory distress, and congenital anomalies, which collectively contribute to over 40% of early neonatal mortality in low- and middle-income countries. Effective neonatal stabilization—encompassing prompt recognition of high-risk conditions, adherence to evidence-based resuscitation protocols, and early supportive care—is fundamental to reducing preventable deaths and long-term morbidity (Biban et al., 2021; Hibbs & Bell, 2015; Wyckoff et al., 2022). However, the complexity of stabilization increases substantially when rare congenital syndromes with multisystem involvement, such as skeletal dysplasias, coincide with acute perinatal complications like meconium aspiration syndrome (MAS) (Dini et al., 2024; Monfredini et al., 2021; Osman et al., 2023). In such scenarios, the intersection of anatomical anomalies and acute respiratory compromise demands heightened clinical vigilance, specialized knowledge, and a multidisciplinary approach to optimize outcomes (Čalkovská et al., 2019; Olicker et al., 2021).

Larsen syndrome is a rare congenital skeletal dysplasia first described by Larsen et al. in 1950, characterized by multiple congenital dislocations of large joints, distinctive craniofacial features, and variable degrees of cervical spine anomalies. The estimated prevalence is approximately 1 in 100,000 live births (Waters et al., 2018; Yu et al., 2023). Affected neonates often present with congenital knee dislocation, cervical spine instability, and

facial features including hypertelorism, a depressed nasal bridge, and a prominent forehead. These anatomical abnormalities present unique challenges during neonatal resuscitation, particularly in airway management and limb positioning (Palco et al., 2022; Siafaka et al., 2023). Unlike resuscitation in healthy neonates, infants with Larsen syndrome require meticulous handling of the head and neck to avoid hyperextension or flexion, which can exacerbate cervical spine instability and potentially cause spinal cord injury (Perlman et al., 2015; Siafaka et al., 2023). During positive pressure ventilation or intubation, the airway should be managed in a neutral position with minimal manipulation (Perlman et al., 2015; Siafaka et al., 2023).

The provision of neonatal care in peripheral or resource-limited hospital settings presents additional challenges that significantly impact stabilization outcomes. Peripheral hospitals often lack advanced diagnostic imaging (such as real-time ultrasound or computed tomography), specialized neonatal intensive care equipment (including conventional mechanical ventilators and surfactant therapy), and immediate access to pediatric subspecialists including neonatologists, pediatric surgeons, and geneticists (Bhutta et al., 2014; Moxon et al., 2015). Furthermore, healthcare providers in these settings may have limited exposure to rare congenital syndromes and complex resuscitation scenarios, necessitating greater reliance on structured protocols, clinical judgment, and timely referral networks (Wall et al., 2010). The combination of limited resources and high-risk clinical presentations underscores the critical importance of evidence-based, protocol-driven care and effective communication among interdisciplinary teams to ensure patient safety and optimize outcomes (Ersdal et al., 2013).

Additionally, the presence of meconium aspiration syndrome further complicates stabilization due to the risk of severe respiratory compromise and hypoxemia (Čalkovská et al., 2019; Olicker et al., 2021). MAS occurs in approximately 5-10% of deliveries complicated by meconium-stained amniotic fluid and is associated with significant respiratory morbidity, including persistent pulmonary hypertension, pneumothorax, and prolonged mechanical ventilation. The pathophysiology involves mechanical airway obstruction, chemical pneumonitis, and surfactant dysfunction, all of which contribute to ventilation-perfusion mismatch and hypoxemia (Alweshahi et al., 2025; Kerr & Neto, 2024; Powers & Dhamoon, 2021; Slobod et al., 2022). When MAS coexists with congenital anomalies that compromise airway anatomy or respiratory mechanics, the risk of adverse outcomes is substantially amplified, requiring careful integration of resuscitation principles with syndrome-specific considerations (Gupta, 2017).

In resource-limited settings, where advanced airway equipment and imaging are often unavailable, awareness of these considerations and adherence to structured resuscitation protocols are essential to minimize morbidity and optimize outcomes (Perlman et al., 2015; Siafaka et al., 2023). Despite the well-established global burden of neonatal complications and the recognized challenges of managing rare syndromes in peripheral settings, there remains a paucity of published case reports and clinical guidance specifically addressing the stabilization of neonates with Larsen syndrome complicated by MAS in resource-constrained environments. This gap in the literature limits the dissemination of practical strategies and evidence-based protocols that could inform clinical decision-making in similar scenarios (Siafaka et al., 2023).

The urgency of this case report is underscored by several critical factors. First, the rarity of Larsen syndrome (prevalence approximately 1 in 100,000 live births) means that most clinicians, particularly those practicing in peripheral hospitals, may encounter this condition only once or twice in their careers, if at all. This limited exposure increases the risk of delayed recognition, inappropriate management, and iatrogenic injury during resuscitation. Second, the concurrent presence of MAS—a relatively common complication of meconium-stained deliveries—in a neonate with underlying skeletal dysplasia creates a compound risk scenario that demands immediate, expert-level decision-making often without the benefit of subspecialty consultation or advanced imaging. Third, errors in airway management or limb handling during the critical "golden minute" of resuscitation can result in catastrophic outcomes, including spinal cord injury, failed ventilation, and death (Perlman et al., 2015; Chen, n.d.).

The novelty of this case report lies in its documentation of successful neonatal stabilization strategies in a peripheral hospital setting where resources were limited but adherence to structured resuscitation protocols and clinical vigilance enabled a favorable outcome. To our knowledge, this is among the first detailed reports from a resource-limited setting that explicitly outlines the stepwise approach to managing a neonate with suspected Larsen syndrome and MAS, emphasizing practical modifications to standard resuscitation algorithms. Furthermore, this report contributes to the growing body of evidence supporting the feasibility and effectiveness of non-invasive respiratory support as a first-line intervention in neonates with complex respiratory and skeletal conditions, thereby reducing the need for endotracheal intubation and its associated risks.

The primary objective of this case report is to describe the clinical presentation, initial stabilization strategies, and early management of a term neonate with suspected Larsen syndrome complicated by MAS in a peripheral hospital setting. Specific aims include: (1) detailing the resuscitation approach with emphasis on airway management considerations specific to neonates with cervical spine instability; (2) illustrating the role of non-invasive positive pressure ventilation in managing respiratory compromise without the need for invasive mechanical ventilation; (3) highlighting the importance of multidisciplinary collaboration among neonatology, orthopedics, and nursing teams in resource-limited environments; and (4) providing practical clinical guidance that may inform the management of similar cases in comparable settings.

The benefits of this case report are multifaceted. From a clinical perspective, it offers a roadmap for healthcare providers in peripheral hospitals who may encounter rare congenital syndromes with limited resources and subspecialty support. By demonstrating that successful outcomes are achievable through adherence to structured protocols and careful clinical assessment, this report may enhance confidence and competence among frontline clinicians. From an educational standpoint, the case provides valuable teaching material for medical students, residents, and practicing clinicians in neonatology and pediatrics. The implications for clinical practice include the reinforcement of protocol-driven care, the importance of early recognition of congenital anomalies, and the value of conservative, non-invasive management strategies when advanced interventions are not immediately available. Ultimately, this report contributes to the broader goal of reducing preventable neonatal morbidity and mortality by

disseminating actionable knowledge and best practices for managing complex clinical scenarios in diverse healthcare settings.

RESEARCH METHODS

This case report describes the clinical course of a term female neonate delivered at a peripheral hospital in Indonesia. The case documentation was conducted in accordance with ethical principles outlined in the Declaration of Helsinki and adheres to the CARE (Case Report) guidelines for transparent reporting of clinical cases. Informed consent was obtained from the patient's parents for the publication of clinical data and de-identified photographs for educational and scientific purposes. The study was approved by the hospital's ethics review committee.

RESULTS AND DISCUSSION

A term female neonate was delivered spontaneously in the delivery room at 15:17. The pregnancy had reached 40 weeks of gestation and was complicated by intrauterine growth restriction and suspected intrauterine infection. The amniotic fluid was thickly stained with meconium. Immediately after birth, the infant did not cry and appeared hypotonic and cyanotic. Initial resuscitation steps, including airway clearance and stimulation, were performed, followed by positive pressure ventilation to facilitate lung aeration. The resuscitation process was conducted in accordance with the 2022 neonatal resuscitation guidelines issued by the Indonesian Pediatric Society (Ikatan Dokter Anak Indonesia/IDAI). Within a few minutes, the infant began to cry, and clinical color improved. Apgar scores were recorded as 3 at one minute, 4 at five minutes, and 5 at ten minutes.

After stabilization, the infant was transferred to the neonatal intensive care unit. On admission, the baby appeared clinically stable with a heart rate of 168 beats per minute, respiratory rate of 63 breaths per minute, and temperature of 36.5°C. Initial oxygen saturation was 93%, which gradually increased to 95–98% after resuscitation. Anthropometric measurements revealed a birth weight of 2200 grams, length of 43 cm, head circumference of 31 cm, chest circumference of 29 cm, and mid-upper arm circumference of 9 cm, consistent with intrauterine growth restriction. Physical examination noted a positive cephalhematoma but no caput succedaneum. The thorax was symmetric, with vesicular breath sounds and no retractions or adventitious sounds. Cardiovascular examination was normal, and the abdomen was soft and flat with normal peristalsis. Examination of the extremities revealed bilateral congenital knee dislocation with marked flexion deformities, as well as fixed flexion deformities of both elbows.

Laboratory investigations showed a hemoglobin of 15.1 g/dL, leukocytosis with a white blood cell count of 23,900/uL, and an elevated C-reactive protein of 8.4 mg/L, supporting a risk of early-onset neonatal infection. Serum calcium was mildly reduced at 0.98 mmol/L. Thyroid stimulating hormone was slightly elevated at 38.36 mIU/L, warranting monitoring. A skeletal survey confirmed bilateral congenital knee dislocation with abnormal flexion and overlapping articular surfaces, without evidence of fractures or gross bony dysplasia of the spine or thorax. Respiratory support was initiated using non-invasive positive pressure ventilation (NIPPV) with settings of PIP 15 cmH_2O, PEEP 5 cmH_2O, and FiO_2 40%. Empirical first-line antibiotic therapy was commenced to address the risk of early-onset sepsis. The infant remained stable

without retractions, and oxygenation was maintained. After three days of NIPPV, the baby was transitioned to high-flow nasal cannula without supplemental oxygen. Orthopedic management included application of bilateral lower limb casts to maintain joint alignment. Enteral feeding with breast milk was initiated and well tolerated. After five days of hospitalization, the infant remained clinically stable, showed no signs of respiratory distress, and demonstrated adequate oral intake. The baby was discharged home in good condition, with plans for follow-up with pediatric orthopedics and endocrinology to monitor joint development and thyroid function.

Figure 1. Congenital Knees Dislocation

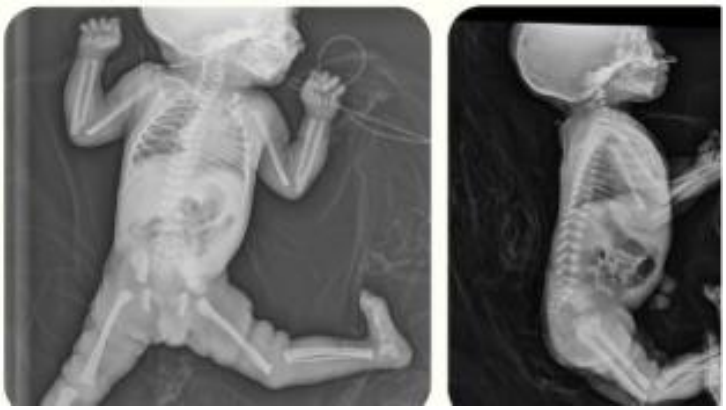

Figure 2. Babygram and Spine X-Ray

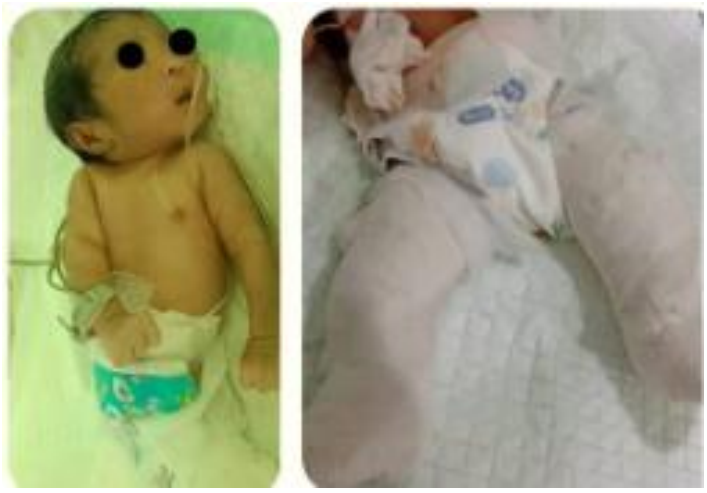

Figure 3. Long Leg Cast Immobilization

Discussions

Larsen syndrome is a rare congenital skeletal dysplasia characterized by multiple joint dislocations, particularly of the knees, hips, and elbows, as well as distinctive craniofacial features such as hypertelorism, a depressed nasal bridge, and a prominent forehead. The syndrome is most commonly caused by autosomal dominant mutations in the FLNB gene, which encodes filamin B, a cytoskeletal protein critical for skeletal development and joint stability. In this case, the presence of bilateral congenital knee dislocation without other major skeletal anomalies supported a provisional diagnosis of Larsen syndrome, pending further genetic evaluation.

Neonates with Larsen syndrome may have cervical spine anomalies, including atlantoaxial instability, which can significantly complicate airway management and resuscitation. Excessive neck extension or flexion during resuscitation maneuvers may result in spinal cord injury. Therefore, careful attention to neck positioning and minimal manipulation are recommended. In this case, resuscitation followed the 2022 neonatal resuscitation guidelines from the Indonesian Pediatric Society (IDAI), emphasizing a neutral head position, airway clearance, and prompt positive pressure ventilation. The timely implementation of these protocols facilitated effective lung aeration and stabilization of the infant's oxygen saturation.

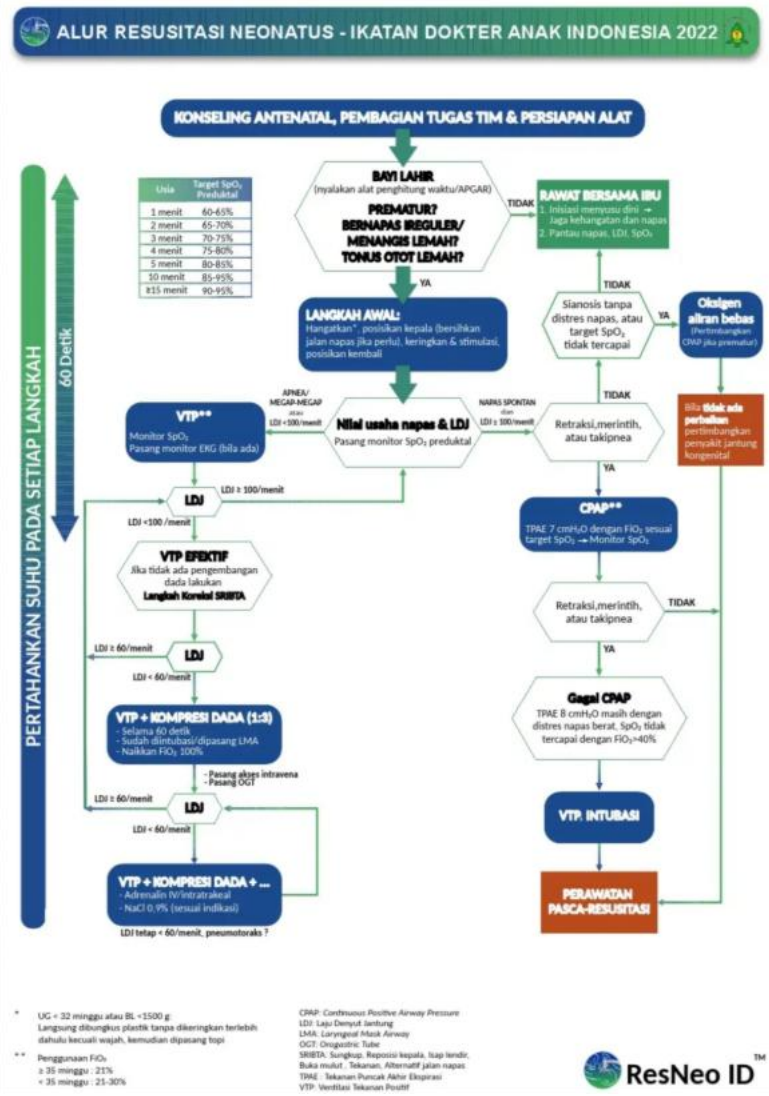

Figure 4. The 2022 neonatal resuscitation guidelines from the Indonesian Pediatric Society (IDAI)

The concurrent presence of thick meconium-stained amniotic fluid and severe respiratory depression at birth raised concerns for meconium aspiration syndrome (MAS). MAS remains a significant cause of neonatal respiratory morbidity and mortality, particularly in resource-limited settings where advanced ventilation strategies may not be readily available. Non-invasive positive pressure ventilation proved effective in maintaining oxygenation without the need for endotracheal intubation or mechanical ventilation. The gradual transition to high-flow nasal cannula and eventual weaning off supplemental oxygen reflected clinical improvement consistent with mild to moderate MAS.

Leukocytosis and elevated C-reactive protein suggested early-onset neonatal sepsis. In neonates exposed to meconium-stained fluid, the risk of infection is increased due to potential intrauterine contamination and aspiration of pathogens. Empirical antibiotic therapy was initiated as per protocol. Additionally, mild hypocalcemia and elevated thyroid-stimulating hormone were identified, warranting monitoring to prevent metabolic complications.

Orthopedic management of congenital knee dislocation is essential to optimize functional outcomes. Early application of gentle traction and serial casting is the mainstay of

treatment and is often effective in achieving joint reduction without surgical intervention. This case underscores the importance of a multidisciplinary approach to neonates with syndromic features and respiratory compromise. Coordination between neonatologists, orthopedic surgeons, and nursing staff is critical to ensure safe stabilization and to minimize the risk of iatrogenic injury. Structured protocols and teamwork can optimize outcomes even in peripheral hospitals.

The multidisciplinary care model employed in this case exemplifies the principles of integrated, patient-centered care. Daily multidisciplinary rounds involving neonatology, orthopedics, nursing, and, when available via telemedicine, genetics and pediatric subspecialists, ensured that all aspects of the infant's care were coordinated and that potential complications were anticipated and mitigated proactively. Communication among team members was facilitated through structured handoffs and shared electronic medical records, which enhanced continuity of care and reduced the likelihood of medical errors.

Furthermore, the role of nursing staff in monitoring, positioning, and providing developmental care cannot be overstated. Nurses were instrumental in maintaining neutral cervical spine positioning during routine care activities, recognizing early signs of respiratory distress, and ensuring adherence to feeding protocols. Parental involvement and education were also prioritized, with clear communication regarding the infant's diagnosis, treatment plan, and expected trajectory, thereby fostering trust and engagement in the care process.

Study Limitations and Future Directions

While this case report provides valuable insights into the management of a neonate with Larsen syndrome and MAS in a resource-limited setting, several limitations must be acknowledged. First, as a single case study, the findings are descriptive and cannot be generalized to all neonates with similar presentations. The absence of comparative data or statistical analysis limits the strength of evidence regarding the efficacy of specific interventions. Second, definitive genetic confirmation of Larsen syndrome via FLNB mutation analysis was not available at the time of discharge, precluding absolute diagnostic certainty. However, the clinical phenotype was highly consistent with the syndrome, and genetic testing was recommended for future follow-up. Third, long-term neurodevelopmental and orthopedic outcomes were not assessed within the scope of this report, as the infant was discharged after only five days of hospitalization. Prospective longitudinal follow-up is essential to evaluate the durability of orthopedic interventions, assess for developmental delays, and monitor for potential complications such as progressive cervical spine instability or recurrent joint dislocations.

Future research should focus on establishing evidence-based protocols for the recognition and initial management of rare congenital syndromes in peripheral hospital settings. Multicenter case series or registry-based studies could provide a larger dataset to inform clinical decision-making and identify predictors of favorable outcomes. Additionally, the role of telemedicine in providing real-time subspecialty consultation to remote facilities warrants further exploration, as it may enhance diagnostic accuracy, guide management, and facilitate timely referrals when necessary. Finally, educational initiatives targeting healthcare providers in resource-limited settings should emphasize the recognition of rare syndromes,

adherence to structured resuscitation protocols, and the principles of gentle, non-invasive neonatal care to improve outcomes for vulnerable infants.

CONCLUSION

This case illustrates the complex challenges associated with the resuscitation andstabilization of neonates with suspected Larsen syndrome complicated by meconium aspiration syndrome and intrauterine growth restriction. Careful attention to airway management—including maintaining a neutral head position to protect the cervical spine—and adherence to structured resuscitation guidelines were critical to achieving effective ventilation without causing additional injury. Early initiation of non-invasive respiratory support, empirical antibiotic therapy, and prompt orthopedic intervention contributed to a favorable outcome. A multidisciplinary approach and close monitoring allowed the infant to recover clinically and be safely discharged in good condition.

REFERENCES

Alweshahi, R., Elwafi, M., Mtm, A., Azab, Abdulmola, K., & Aaa, M. (2025). A Comprehensive Review of Respiratory Failure: Pathogenesis, Clinical Strategies, Ethical Challenges, and Preventive Approaches: A Literature Review. *Libyan Journal of Medical Research*. https://doi.org/10.54361/LJMR.19.2.28

Amuka, J. I., Asogwa, F. O., Ugwuanyi, R. O., & Onyechi, T. (2020). Neonatal deaths and challenges of public health: Where do We need urgent intervention in developing countries? *Health Care for Women International*.

Biban, P., Marlow, N., Pas, A. B. Te, Fanaroff, A. A., & Jobe, A. H. (2021). *Advances in Neonatal Critical Care: Pushing at the Boundaries and Connecting to Long-Term Outcomes.*

Čalkovská, A., Mokrá, D., Čalkovský, V., Matasová, K., & Zibolen, M. (2019). Clinical considerations when treating neonatal aspiration syndromes. *Expert Review of Respiratory Medicine*. https://doi.org/10.1080/17476348.2019.1562340

Dini, G., Ceccarelli, S., Celi, F., Semeraro, C. M., Gorello, P., & Verrotti, A. (2024). Meconium aspiration syndrome: from pathophysiology to treatment. *Annals of Medicine and Surgery*. https://doi.org/10.1097/MS9.0000000000001835

Guerrera, G. (2015). Neonatal and pediatric healthcare worldwide: A report from UNICEF. *Clinica Chimica Acta*.

Gupta, P. (2017). *Translating Mouse Systems Genetics to Discovery in Human Disease*.

Hibbs, B., & Bell, M. A. (2015). Collaborating to Create a Comprehensive Neonatal Resuscitation Form. *Journal of Obstetric, Gynecologic, & Neonatal Nursing*.

Hug, L., Alexander, M., You, D., & Alkema, L. (2019). National, regional, and global levels and trends in neonatal mortality between 1990 and 2017, with scenario-based projections to 2030: a systematic analysis. *The Lancet Global Health*.

Kerr, C. L., & Neto, F. J. T. (2024). *Respiratory Physiology and Pathophysiology*. https://doi.org/10.1002/9781119830306.CH37

Monfredini, C., Cavallin, F., Villani, P., Paterlini, G., Allais, B., & Trevisanuto, D. (2021). Meconium Aspiration Syndrome: A Narrative Review. *Children Today*. https://doi.org/10.3390/CHILDREN8030230

Moradi-Lakeh, I. M. C. R. M. A. K. J. K. C. E. B. C., Khalil, I., Collison, M. L., Bcheraoui, C. El, Charara, R., Moradi-Lakeh, M., Afshin, A., Krohn, K. J., Daoud, F., Chew, A., Kassebaum, N., Colombara, D., Cornaby, L., Ehrenkranz, R., Foreman, K., Fraser, M. S., Frostad, J., Kemmer, L., Kulikoff, X., … Mokdad, A. (2018). Neonatal, infant, and

under-5 mortality and morbidity burden in the Eastern Mediterranean region: findings from the Global Burden of Disease 2015 study. *International Journal of Public Health*.

Olicker, A. L., Raffay, T. M., & Ryan, R. M. (2021). Neonatal Respiratory Distress Secondary to Meconium Aspiration Syndrome. *Children Today*. https://doi.org/10.3390/CHILDREN8030246

Osman, A., Halling, C., Crume, M., Tabosh, H. Al, Odackal, N. J., & Ball, M. K. (2023). Meconium aspiration syndrome: a comprehensive review. *Journal of Perinatology*. https://doi.org/10.1038/S41372-023-01708-2

Powers, K. A., & Dhamoon, A. S. (2021). Physiology, Pulmonary Ventilation and Perfusion. *StatPearls*.

Slobod, D., Damia, A., Leali, M., Spinelli, E., & Mauri, T. (2022). Pathophysiology and Clinical Meaning of Ventilation-Perfusion Mismatch in the Acute Respiratory Distress Syndrome. *Biology*. https://doi.org/10.3390/BIOLOGY12010067

Tessema, G., Berheto, T., Pereira, G., Misganaw, A., & Kinfu, Y. (2023). National and subnational burden of under-5, infant, and neonatal mortality in Ethiopia, 1990–2019: Findings from the Global Burden of Disease Study 2019. *PLOS Global Public Health*.

Waters, D., Adeloye, D., Woolham, D., Wastnedge, E., Patel, S., & Rudan, I. (2018). Global birth prevalence and mortality from inborn errors of metabolism: a systematic analysis of the evidence. *Journal of Global Health*. https://doi.org/10.7189/JOGH.08.021102

Wyckoff, M. H., Singletary, E. M., Soar, J., Olasveengen, T. M., Greif, R., Liley, H. G., Zideman, D., Bhanji, F., Andersen, L. W., Avis, S. R., Aziz, K., Bendall, J. C., Berry, D. C., Vere Borra, Böttiger, B. W., Bradley, R., Bray, J. E., Breckwoldt, J., Carlson, J. N., … Berg, K. M. (2022). 2021 International Consensus on Cardiopulmonary Resuscitation and Emergency Cardiovascular Care Science With Treatment Recommendations: Summary From the Basic Life Support; Advanced Life Support; Neonatal Life Support; Education, Implementation, and Teams; First Aid Task Forces; and the COVID-19 Working Group. *Circulation*.

Yu, D., Ling, L. E., Krumme, A., Tjoa, M. L., & Moise, K. J. (2023). Live birth prevalence of hemolytic disease of the fetus and newborn in the United States from 1996 to 2010. *AJOG Global Reports*. https://doi.org/10.1016/J.XAGR.2023.100203