

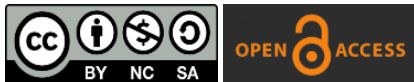
Prenatal Diagnosis of Prune Belly Syndrome and Potter Sequence: A Complex Clinical and Ethical Scenario

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Received: September 09, 2023; Accepted: October 16, 2023; Published: October 29, 2023



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Abstract

Prune-belly syndrome, or Eagle-Barrett syndrome, is a rare congenital anomaly characterized by genitourinary abnormalities and aplastic or hypoplastic abdominal muscles. We present a case of prenatal diagnosis of prune belly syndrome at 20 weeks gestation, complicated by Potter sequence due to obstructive uropathy. Despite counseling, restrictive abortion laws limited the mother's options, leading to a continued pregnancy with close monitoring and palliative care. The infant exhibited severe features of Potter sequence and succumbed shortly after birth. This case underscores the challenges parents face in restrictive abortion law environments and emphasizes the importance of comprehensive prenatal counseling and access to medical services, including termination when medically indicated.

Keywords: Prune belly syndrome; Potter sequence; Neonatal complications.

1. Introduction

Prune-belly syndrome, also known as Eagle-Barrett syndrome, is a rare congenital anomaly characterized by severe genitourinary tract abnormalities in combination with aplastic or hypoplastic abdominal musculature [1]. In some fetuses, urethral obstruction may lead to severe oligohydramnios with resulting Potter sequence. Thus, Potter sequence and Prune-belly syndrome may coexist when dysfunctional kidneys cause an obstructive uropathy resulting in severe oligohydramnios [2]. We present a case of a baby diagnosed with prune belly at 20 weeks gestation. It is likely that this patient had obstructive uropathy secondary to Prune-belly syndrome, which led to subsequent Potter sequence. Common features of Potter sequence include pulmonary hypoplasia, limb deformities (e.g., clubbed feet), and characteristic facial anomalies [3]. The baby was delivered via cesarean section at 34 weeks and died at birth.

Citation: Mishra A and Marcuzzo G. Prenatal diagnosis of prune belly syndrome and potter sequence: A complex clinical and ethical scenario. Case Rep Rev Open Access. 2023;4(2):133.

2. Case Presentation

A woman presented for a routine prenatal ultrasound at 20 weeks gestation. During the ultrasound, significant findings included bilateral hydronephrosis with hydroureter, distension of the fetal abdomen, and absence of discernible abdominal wall musculature. The parents were counseled about the implications on fetal development, including the risk of Potter sequence.

Despite the diagnosis, the mother faced significant barriers to accessing care. Nebraska's abortion laws limited her options, and she could not afford to travel out of state for termination. Consequently, the pregnancy continued, and the patient was closely monitored for complications related to prune-belly. She was followed by palliative care. Serial ultrasounds revealed progressive oligohydramnios, reduced fetal lung development, and limb deformities consistent with Potter sequence. The baby was delivered by elective C-section at 34 weeks gestation to optimize neonatal care.

The newborn male infant exhibited typical features of Potter sequence, including severe pulmonary hypoplasia, limb deformities, a flat face with a broad nasal bridge, prune belly, and prominent epicanthal folds. Despite aggressive resuscitative efforts in the neonatal intensive care unit (NICU), the infant's respiratory distress was refractory to treatment, and he sadly succumbed shortly after birth.

3. Discussion

Prune Belly Syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare congenital anomaly characterized by a complex triad of abdominal wall muscle deficiency, cryptorchidism, and genitourinary tract abnormalities [1]. In this case, the prenatal diagnosis of PBS at 20 weeks gestation brought to light not only the clinical complexities of managing this syndrome but also the intricate ethical and legal challenges associated with restrictive abortion laws.

The presence of bilateral hydronephrosis with hydroureter, distension of the fetal abdomen, and absence of discernible abdominal wall musculature strongly indicated PBS. Furthermore, the risk of Potter sequence was explained to the parents due to obstructive uropathy caused by PBS [3]. Potter sequence encompasses a spectrum of severe abnormalities, including pulmonary hypoplasia, limb deformities, and distinct facial anomalies, all of which were observed in the newborn [4].

Despite comprehensive counseling regarding the grave implications of the diagnoses, the mother in this case faced significant obstacles in accessing appropriate medical care. Stringent abortion laws in the state limited her options, preventing her from pursuing termination, which is often considered the most common therapy for antenatal bladder outlet obstruction. The neonate's clinical presentation after birth was consistent with the anticipated severe features of Potter sequence. Despite the dedicated efforts of the neonatal intensive care unit, the infant's respiratory distress proved refractory to treatment, and the baby passed away shortly after birth.

4. Conclusion

This case underscores the intricate interplay of medical, ethical, and legal factors when confronted with a prenatal diagnosis of Prune Belly Syndrome and its potential complications, such as Potter sequence. The challenges faced by the parents in a restrictive abortion law environment exemplify the difficulties that some families encounter in accessing comprehensive prenatal care and the option of termination when medically indicated.

It is imperative for healthcare systems and legislators to recognize the complexities surrounding cases like this one and to ensure that expectant parents receive comprehensive prenatal counseling, education, and access to appropriate medical services, including the option of termination when medically indicated. A multidisciplinary approach, including genetic counselors, neonatologists, and maternal-fetal medicine specialists, is essential in navigating these challenging scenarios.

The case also highlights the need for continued research and advocacy to raise awareness about PBS and related conditions, fostering a better understanding of their management and the challenges faced by affected families. Ultimately, comprehensive and compassionate care remains paramount in providing the best possible outcomes for both mothers and infants in such complex and emotionally charged situations.

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