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Case Report

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Labor in a Woman With Prune Belly Syndrome

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Abstract

Pregnancy and labor in patients with Prune Belly syndrome (PBS) have only been described in three cases in the literature. In these case reports, two patients had elective cesarean section due to PBS and consequences of the syndrome. One patient attempted vaginal delivery, had a prolonged second stage of labor and delivered by vacuum-assisted delivery. We aimed to investigate the feasibility of vaginal delivery in women with hypoplastic abdominal muscles without the need for instrumental or operative intervention. We report a patient with PBS, only affecting the abdominal muscles, who attempted vaginal delivery. Due to prolonged second stage of labor and exhaustion and after being fully dilated for 3 h with no evident progression, an emergency cesarean section was performed. The second stage of labor involves coordinated uterine contractions and voluntary maternal efforts. Impairment of the second stage of labor can occur if the woman is unable to effectively push, due to factors such as hypoplastic abdominal muscles. This inability to generate sufficient intra-abdominal pressure can lead to prolonged labor, increased risk of fetal distress, and the potential need for instrumental delivery or cesarean section to ensure the safety of both the mother and the fetus. Our case supports the thesis that patients with PBS may experience prolonged second stage of labor and need assisted instrumental or operative delivery. Whether it is possible for a patient with PBS to deliver vaginally without instrumental or operative interference is yet unknown.

Keywords: Prune Belly syndrome; Eagle-Barrett syndrome; Prolonged second stage of labor

Introduction

Prune Belly syndrome (PBS), also known as Eagle-Barrett syndrome, is a rare and complex congenital disorder with an incidence of 1 in 29,000 to 40,000 live births, with 95% reported in males [1], characterized by the classic triad of abdominal wall muscular deficiency, resulting in prune-like abdominal wall, bilateral cryptorchidism, and dilated urinary tract in men. PBS in women presents with abdominal wall muscular deficiency and urinary tract abnormalities, but with no gonadal anomalies. The severity of the condition ranges from stillbirth to significant abnormalities to almost normal patients and the prognosis mainly depends on the degree of lung and renal abnormalities [2]. The etiology of the condition is yet unknown but there are two prevailing theories. One is that an early *in utero* posterior urethral obstruction results in dilation of the urinary tract, possible fetal ascites and oligohydramnios, although this does not explain the manifestations outside the genitourinary system [3]. The other theory suggests that altered gene regulation of myogenesis causes maldevelopment of the somatic and splanchnic mesoderm resulting in dysplastic abdominal wall muscles and urinary tract malformation [4].

In the literature, there are only three reported cases of pregnancy and labor in women with PBS. In the case report by Hillman et al [5], a woman with PBS attempted vaginal delivery. In childhood, she had suffered from several urinary infections and recurrent pneumonia, but there were no detected urinary tract abnormalities. There was only evidence of PBS affecting her abdominal muscles. Pregnancy was uncomplicated and delivery was accomplished with vacuum-assisted delivery, due to prolonged second stage of labor. Morita et al reported a case of a woman diagnosed with PBS due to fetal hydrops and abdominal wall hypoplasia [6]. In childhood, she underwent open bowel resection due to an intestinal malrotation and funnel chest surgery. She suffered from abdominal organ retention caused by abdominal wall dysplasia. She was diagnosed with primary infertility and her pregnancy was achieved using fertility drugs. Pregnancy was complicated by PBS due to the severity of hypoplasia of the abdominal wall which was very thin and flaccid. The abdominal wall was displaced anteriorly and the pregnant uterus could be seen just below the abdominal wall. Magnetic resonance imaging was performed and showed that the rectus abdominis muscle was thinning and the oblique abdominal muscle was hardly visible. The patient was recommended a cesarean section due to concerns about prolonged second stage of labor, and she accepted. The surgery was without noticeable complications, confirming that the abdominal wall was significantly thinner than usual. In the case report by Moreno et al [7], a patient with PBS had no abdominal muscles and suffered from asthma and severe kyphoscoliosis. She had no urinary tract malformations. The pregnancy was complicated by compression of the abdominal organs, causing frequent constipation, abdominal bloating and epigastric pain. She gained only 3.4 kg during pregnancy.

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Ultrasonography revealed possible intrauterine growth restriction with an estimated fetal weight below the third percentile. Due to abnormal maternal pelvic rotation and transverse position of the fetus and possible intrauterine growth restriction, elective cesarean section was performed at 37 weeks of gestation. During surgery, the absence of abdominal muscles and typical aponeurosis was noted, but the surgery was without complications.

In this article, we report a case of a women with PBS resulting in hypoplastic abdominal muscles, who attempted vaginal delivery, but ended up having an emergency cesarean section due to prolonged second stage of labor.

Case Report

Investigations

A 29-year-old pregnant woman with PBS was referred from the routine prenatal care unit to the obstetric clinic in week 12 of gestation in order to do extra checkups and decide on a birth plan during the pregnancy. Immediately after she was born, she was admitted at the pediatric department due to hypoplasia of the abdominal muscles. Further investigations revealed no abnormalities in the urinary tract or intestines. She was diagnosed with PBS that affected only her abdominal muscles which were hypoplastic. During childhood, she was diagnosed with asthma, irritable bowel syndrome and attention deficit hyperactivity disorder. She did not take any medication, and worked full time as a social and health care assistant. The PBS did not result in any severe restrictions in her everyday life.

Diagnosis

At the time of referral, the patient was gravida 2, para 0, and had a history of a surgical induced abortion, at gestational age 8 + 3, 5 years prior to this pregnancy. The pregnancy was planned and there were no reported incidences of infertility or conception difficulties. Before the pregnancy, she weighed 50 kg, was 163 cm, with a body mass index of 18.8. During the first trimester, she suffered from nausea and vomiting, which was treated with vitamin B6. She experienced light vaginal bleeding several times during the first trimester, but was only in contact with her general practitioner and not admitted to the hospital. No treatment for vaginal bleeding was initiated and the bleeding stopped after the first trimester. From 20 weeks of gestation, she started having lower back pain, which worsened throughout the pregnancy. The pain was treated with trochanter belt with sparse effect and she went on sick leave from work until maternity leave. During the pregnancy, regular ultrasonography was performed in the weeks 12, 20, 28 and after this every fourth week until delivery. The ultrasonography revealed normal development and weight gain of the fetus. The possibility of vaginal birth or planned cesarean section was discussed with the patient, explaining that the hypoplastic abdominal muscles could cause an insufficient increase in abdominal pressure, leading to prolonged second stage of labor. The existing literature of labor in patients with PBS was presented for the patient and her spouse. After thorough information, the patient wished to pursue vaginal birth, knowing that there was a risk of conversion to acute cesarean section or vacuum-assisted delivery due to insufficient progression in labor.

Treatment

At 39 + 1 weeks of gestation, she had spontaneous rupture of membranes but there were no spontaneous contractions. After 14 h with no contractions, she received misoprostol to try to induce labor. The estimated weight of the fetus was 3,400 g. We tested group B streptococcus status, which was negative. She started having contractions, but they were ineffective and irregular, she was 2 cm dilated when the contractions started and only 3 cm dilated after having irregular painful contractions for 10 h. After epidural anesthesia was initiated, we started stimulation with oxytocine under continuous cardiotocography-monitoring. During stimulation with oxytocine, she went from being 4 cm dilated to fully dilated within 5 h. When she had been fully dilated for 3 h, the caput was at the level of ischial spines and the patient was exhausted. She was given the option of cesarean section or increased stimulation with oxytocine and chose cesarean section.

Follow-up and outcomes

Emergency cesarean section was performed in spinal anesthesia. A low cross-section in the skin incision was made and during the blunt dissection of the layers the abdominal muscles were observed to be very thin. The operation was complicated by uterine atony after delivery, but after supplementing with oxytocine bolus the uterus was well contracted and there was hemostasis. Blood loss during surgery was 850 mL. The newborn had full Apgar score after 1 min.

Discussion

PBS is a rare condition and the majority of PBS cases have been reported in males. In our literature search, we found only three case reports discussing the method of labor in women with PBS. In these case reports, two patients had elective cesarean section [6, 7] and one attempted vaginal delivery, accomplished with vacuum assistance due to “prolonged second stage of labor secondary to diminished maternal pushing forces” [5].

The labor mechanism is a physiological process initiated by hormonal triggers leading to rhythmic uterine contraction and progressive cervical effacement and dilation. The second stage of labor witnesses the descent and engagement of the fetal head within the maternal pelvis, guided by cardinal movements that facilitate its passage through the birth canal. The mother actively participates in this stage through pushing efforts, culminating in the delivery of the fetus.

We report a woman with hypoplastic abdominal muscles as the only manifestation of PBS. She was presented with the

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option to deliver by either planned cesarean section or vaginal delivery and she wished to attempt vaginal delivery. In the first stage of labor, there was a need of supplemental uterine contracting medications to ensure sufficient progression due to prelabor rupture of membranes and this resulted in a full dilation of the cervix. Since the first stage of labor is mainly facilitated by contractions of the uterus, there are no reasons to believe that this stage would be compromised by the lack of abdominal muscles. However, the second stage of labor typically requires the mother to actively contract the abdominal muscles simultaneously with uterine contractions to ensure sufficient intra-abdominal pressure. In this case, the second stage of labor was prolonged with no evident progression after 3 h of being fully dilated. The patient was exhausted and an emergency cesarean section was performed. This could be an indication that hypoplastic abdominal muscles may cause prolonged second stage of labor due to a lack of maternal pushing forces leading to an increased risk of cesarean section during labor. In a case report by Hillman et al [5], it was possible to go through with vaginal delivery despite prolonged second stage of labor by intervening with vacuum-assisted delivery. There are no reports describing labor without instrumental or surgical intervention in a patient with PBS in the existing literature. Whether it is possible for a woman with PBS to deliver vaginally with no instrumental or surgical intervention is yet unknown.

Learning points

This case supports the thesis that patients with PBS are at risk of experiencing prolonged second stage of labor leading to the need of assisted instrumental or operative delivery.

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Conflict of Interest

None to declare.

Informed Consent

The patient has given informed consent.

Author Contributions

Kirsten Bunemann Jacobsen wrote the manuscript with inputs and editing from Richard Farlie.

Data Availability

All data were collected from the patient file. The data are not publicly available due to their containing information that could compromise the privacy of the patient.

Abbreviations

PBS: Prune Belly syndrome

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