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Case Report

Evolution and Obstetric Outcome in An Adolescent With Cystic Fibrosis Monitored by A Reference Center in Brazil:A Case Report - @

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ABSTRACT

Several parameters such as the pre-pregnancy pulmonary status, forced vital capacity, nutritional status, pancreatic insufficiency, diabetes mellitus, frequent respiratory exacerbations and colonisation with *Burkholderiacepacia* have been associated with adverse pregnancy outcomes. Despite of these aspects, careful monitoring by a multidisciplinary team in a reference center contributes to a successful pregnancy. We report successful pregnancy in adolescent with cystic fibrosis in Brazil.

Key words: Cystic fibrosis; Adolescent pregnancy; Nutritional care

INTRODUCTION

Various physiological and clinical parameters such as the pre-pregnancy pulmonary status [Forced Expiratory Volume in 1 second (FEV₁), Forced Vital Capacity (FVC)], nutritional status, pancreatic insufficiency, diabetes mellitus, frequent respiratory exacerbations and colonisation with *Burkholderiacepacia* have been associated with adverse pregnancy outcomes [1, 2].

To our knowledge, there are still no Brazilian studies on nutritional care during pregnancy and the obstetric outcome in women with Cystic Fibrosis (CF). This case report is the first to describe the evolution and obstetric outcome in an adolescent with CF monitored by a Reference Center.

CASE REPORT

The case involves an 18 years old teenager, primigravid and diagnosed with CF, heterozygous for the G542X mutation. In the pre-pregnancy period, she showed normal oral glucose tolerance test (OGTT), pancreatic sufficiency, nutritional adequacy (body mass index for age (BMI/A of 19.83 kg/m²), colonization by non-mucoid *Pseudomonas aeruginosa*, and pulmonary function testing with FEV₁ of 50% and FVC of 82%.

At 14 weeks' gestation, the patient showed pulmonary function testing with 53% FEV₁ and 74% FVC. At 21 weeks' gestation, the nutritional adequacy was maintained (BMI/A of 20.35 kg/m²), she received diet prescription with 4100 kcal of total energy value (TEV), conferred by the diet plus the oral caloric supplement, distributed in 51% of carbohydrate, 18% of protein and 31% of lipids, divided into 06 meals/day. At 31 weeks' gestation, the patient kept the nutritional adequacy, BMI/A of 22.30 kg/m², FEV₁ of 59.7% and FVC of 74%, maintaining the nutritional guidance. There are reports of hospitalization for five days at the University Hospital in the city of Campos due to pneumonia, which was treated with ceftriaxone.

During the prenatal period, the follow drugs were used: mucolytic enzyme (Dornase alfa), emergency bronchodilator (Salbutamol[®]), inhaled corticosteroid (Seretide[®]), vitamin and mineral supplement (Folic Acid and iron). Also, the patient received oral caloric supplement (Sustagem[®] and vegetable oil).

The patient had a total weight gain of 9.5 kg without edema; she reported episodes of nausea and vomiting early in the first trimester of gestation and increased appetite during pregnancy. Showed no changes in blood pressure and glucose. There were no reports of obstetric and perinatal complications. The obstetrical outcome was a living female newborn, pre-term (35 weeks), weighing 2705g, length of 49 cm, head circumference of 34 cm and APGAR scores of 7 and 8.

In the second postpartum month, the patient kept the nutritional adequacy, BMI/A of 20.4 kg/m², despite keeping her daughter under Exclusive Breast Feeding (EBF).

DISCUSSION

This study shows that women with CF with an adequate nutritional status and moderately impaired pulmonary function, if monitored by a multidisciplinary team, have more chances to reach a successful obstetric outcome, which is in agreement with other studies [3, 4].

In the CF, malnutrition is a common, multifactorial finding, and low prepregnancy weight, BMI below 18 kg/m² (<85% ideal body weight) is associated with an unfavorable pregnancy outcome [5]. In this case, the patient remained eutrophic from pre-pregnancy to late gestation, even though the nutritional demand increased by CF, adolescence and pregnancy.

The nutritional handling in pregnant women with CF is based on dietary recommendations for the general population of pregnant women followed by specific nutritional requirements of the CF [5]. It is recommended that the diet daily TEV corresponds to 120-150% of recommended daily intake (RDI) for healthy individuals with an additional 200 to 300 Kcal/day [6]. The daily TEV described in the case was of 4100 kcal, which corresponds to 146% of RDI with 51% carbohydrate, 18% protein and 31% fat, preferably the long-chain triglycerides, to ensure proper intake of essential fatty acid, in particular, docosahexaenoic acid (DHA), essential for the development of the fetal nervous system and presented in decreased levels in CF patients [7].

Weight gain is an essential component for fetal growth and development, and CF patients often encounter problems gaining weight during pregnancy [2]. A total weight gain of at least 10 kilograms during pregnancy in women with CF has been associated with positive obstetric outcomes, and it is similar to the average of 12.5 kg in women without CF [8], striving for favorable outcomes for both mother and fetus health. In fact, the patient studied earned a total weight of 9.5 kg and presented a preterm newborn as an obstetrical outcome.

Although the diabetes related to the CF is a major complication since the adolescence [9] and pregnant women with CF show a higher risk of acquiring gestational diabetes mellitus [10], the patient did not develop this metabolic alteration.

In this case, the spirometry showed that there was no worsening of the lung damage development, probably due the more intensive antibiotic therapy, higher therapeutic arsenal available today in CF care, and improved adherence to treatment during pregnancy. However, the patient required hospitalization at 31 weeks gestation for treatment of pulmonary infection. The moderately severe [11] impairment of this patient's lung function (FEV₁ 53% and 59.7%), may also have favored the preterm birth. As it is known, most complications for the mother are to deteriorating lung function and nutrition and, for the fetus thought to be related to prematurity [12].

For some mothers with CF breastfeeding can be successfully undertaken. However, it is not possible for many women, due to poor milk flows and to the further nutritional demands it places on the mother [13].

In the case reported, the patient kept the two months old daughter in EBF. Many women with CF are unable to keep it for six month, according to the recommended. Consensus is that women should be encouraged to continue the EBF as long as possible. If needed, can supplement it with initial baby formula or follow-up. Close monitoring of nutritional status and fatigue should be undertaken, and consideration for weaning may be desirable for the health of the mother. Thus, it should be monitored if the mother is willing or is physically capable to keep the breastfeeding [14].

Despite the moderately severe impairment of pulmonary function and teenager status, the good nutritional status, absence of diabetes and follow up by a multidisciplinary team in a reference center contributed to satisfactory pregnancy outcome, once the birth occurred at 35 weeks, but the newborn showed appropriate weight for the gestational age. Thus, women with CF can have healthy pregnancies if keeping stable and satisfactory clinical conditions.

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