

Pregnancy in a Woman with Williams-Campbell Syndrome: A Case Report

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Introduction

Williams-Campbell syndrome is a rare congenital disorder characterized by the deficiency of cartilage in subsegmental bronchi, leading to distal airway collapse and diffuse bilateral bronchiectasis [1,2]. There are few reports describing management of gynaecological disease and pregnancy in women affected by this disorder [3]. In particular, there is no data regarding the possible impact of pregnancy on this pulmonary disorder and vice versa. We present a case report of a woman who underwent surgery for leiomyomas and subsequently became pregnant.

Case Report

Since infancy the patient presented with chronic cough and recurrent pneumonia. Cystic fibrosis, primary ciliary dyskinesia and other well known causes of chronic lung disease were ruled out. At 7 yrs of age the diagnosis of Williams Campbell Syndrome was confirmed by bronchography, showing diffuse bronchiectasis and bilateral collapse of the airways during exhalation **Figure 1A**.

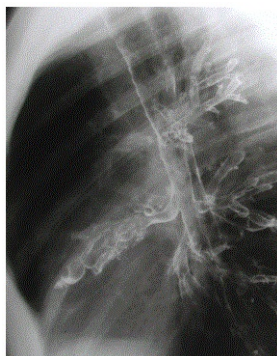


Figure 1A Bronchography during deep inspiration, showing diffuse dilatation of the bronchial tree.

From the age of 6 yrs lung function tests showed severe airflow obstruction, FEV1 ranging 30-40% predicted, and pulmonary hyperinflation. Functional and clinical data remained stable for years. In 1995 chronic lung infection by *Pseudomonas aeruginosa* developed. In 2013 the patient was hospitalized for acute respiratory failure and a diagnosis of tuberculosis was made **Figure 1B**.

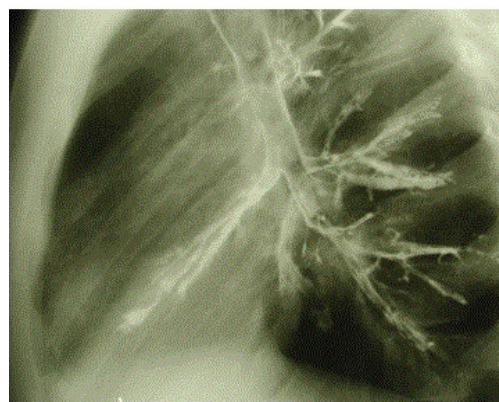


Figure 1B Bronchography showing almost complete collapse of the airways during expiration.

In March 2014, at the age of 35 yrs, she underwent a gynaecological exam for abnormal menstrual bleeding and symptoms indicating growing leiomyomas (diagnosed 2 years prior). At ultrasound examination we found enlarged multiple myomas, at least nine, with a range of 8 mm to 7 cm in diameter. After counselling, we decided to perform a laparotomy, in order to remove all possible myomas, even smaller and intramural.

She was normal weight (56 Kg, body mass index of 21), nulliparous, 2 year use of oral contraceptives; she was taking rifampicin 600 mg once daily dose for recently contracted tuberculosis.

The operation was conducted in balanced general anesthesia. Surgery lasted 85 minutes. Hysteroscopy showed normal uterine cavity. By laparotomy, we removed eight leiomyomas, the largest

was 8 cm diameter. Estimated blood loss was 100 mL. The woman was transferred in Postoperative Intensive Care Unit in stable condition and extubated after 5 hours. Although on antibiotic endovenous therapy (rifampicin 600 mg daily and levofloxacin 500 mg 2 times a day), the postoperative course was complicated by fever thought to be due to airway inflammation a chest X-ray showed the presence of accentuation of lung trama in the right paracardiac region. Continued therapy and care stabilized the patient and she was discharged on the sixth day post-operative.

Pregnancy

In April 2015, 8 months following surgery and at the age of 36 years, the patient became pregnant. First trimester blood tests and ultrasound were normal.

Before pregnancy the patient's pulmonary status was stable, oxygen saturation being 99% while breathing room air, FEV1 1.03 L (35% predicted), and chronic cough and minimal bronchial secretions were the major daily symptoms.

At 23 weeks gestation, during a routine visit, a shortened cervix of 23 mm was detected in the asymptomatic patient. Cardiotocography revealed no uterine contractile activity and blood tests showed no pathological elements. Considering fetal ultrasound estimated weight of 630 g, we undertook corticosteroid treatment (betamethasone 12 mg repeated at 24 hours apart) for accelerating fetal lung maturation.

Urine culture and cervical-vaginal swab were performed; the latter showed the presence of *Candida Spp.* and *Escherichia coli*. Antimicrobial therapy was started with amoxicillin and clavulanic acid (875 mg + 125 mg) 3 times per day for six days.

At the end of October, at 30 weeks gestation, the woman received flu vaccination and corticosteroid profilaxis for fetal lung maturity was repeated due to the appearance of uterine contractions. Through-out pregnancy ultrasound showed regular fetal growth and a normal amount of amniotic fluid **Figure 2**. During pregnancy chest physiotherapy was increased with the purpose to avoid possible pulmonary exacerbations.

Lung function remained stable and no episode of pulmonary exacerbation was recorded.

A caesarean section was chosen for delivery at 39 weeks of gestation, given the history of previous miomectomy and with the aim avoid possible respiratory acute complications during delivery.

Surgery were carried out under L3-L4 subarachnoid anesthesia by 9 mg hyperbaric bupivacaine 0.5%, using a 27G Sprotte needle; antimicrobial prophylaxis with piperacillin was administrated. During surgery O₂ saturation was 98%. Caesarean section lasted 50 minutes, during which time we performed lysis of adhesions between uterus and urinary bladder and removal of keloid cutaneous scar of previous laparotomy. Amniotic fluid was clear and a normal quantity. The blood loss was normal (\approx 500 mL).

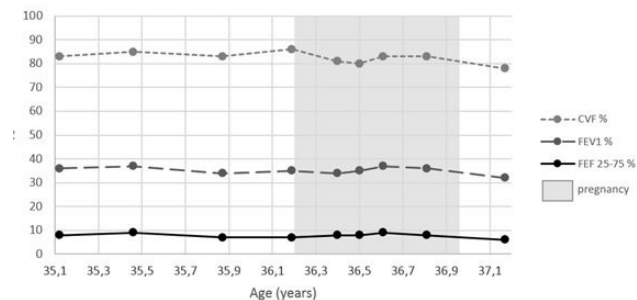


Figure 2 Lung function evolution during pregnancy (grey zone) and in the year before. CVF% =Forced Vital Capacity (% predicted), FEV1%=Forced Expiratory Volume at the first second of CVF (%predicted) and FEF25-75%=Forced Expiratory Flow between 25 and 75% of CVF (% predicted).

The newborn was a male of 3680 g, with Apgar scores of 9/10 and arterial pH of 7.33. The postoperative course was uncomplicated with mother and the child discharged on the fourth day. Pathological examination showed a normal third trimester placenta.

Conclusion

During pregnancy pulmonary function undergoes important changes. Tidal volume, resting minute ventilation and airway conductance increase, while total pulmonary resistance reduces as a result of mechanical and functional factors, including the stimulatory effect of progesterone[4,5]. Moreover, in a patient with severe airflow obstruction a progression towards the development of chronic respiratory failure can be of major concern. Despite such clinical and functional aspects, it seems that in this case the deficiency of cartilage in subsegmental bronchi, that characterizes Williams-Campbell syndrome, did not have any negative effect on lung function during pregnancy nor on fetal growth.

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