CYSTIC FIBROSIS IN PREGNANCY
A DISTRICT GENERAL HOSPITAL EXPERIENCE

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Introduction

Cystic Fibrosis (CF) is an autosomal recessive disorder with an incidence of 1: 2500 live births. Caucasian carrier rate is 1:25. As life expectancy of CF patients are increasing the incidence of pregnancies in CF patients are also increasing. These are very high risk pregnancies with potential risk of high morbidities and mortalities.

We present a case with CF with two pregnancies in our District General Hospital(DGH) where the patient refused to deliver in a tertiary centre.

Case Report

21 years of age, diagnosed with CF at 7 years of age. She has lung disease (baseline FEV1 of 77%), CF related diabetes, pancreatic insufficiency, ulcerative colitis, fixed length gastrostomy tube and Nissen's fundoplication in 1999.

She had 2 successful pregnancies in 2012 and 2016 in a (DGH) in UK. In both her pregnancies, she was managed by joint endocrine antenatal team who co-ordinated care between DGH and tertiary centres.

In her 1st pregnancy, her BMI was 17.5; she was under regular dietician review, on multivitamin supplementation, intermittently requiring feeding via gastrostomy tube which was changed to variable length in pregnancy. Her diabetes was well controlled on insulin. She was on Flucloxacillin throughout her pregnancy for her chest and had regular chest physiotherapy.

She was delivered by emergency caesarean section at 30+ weeks for acute relapse of ulcerative colitis followed by suspicious CTG. Baby's birth weight was 1920 gm and was in good condition at birth, was discharged after 3 weeks.

Post operatively she was transferred to tertiary centre due to lack of respiratory physiotherapy and cystic fibrosis dietary support. She was discharged from the tertiary centre after 3 days.

Case Report(contd.)

In her second pregnancy her CF was stable. She required steroids and Vedolizumab infusion for ulcerative colitis. Her diabetes was managed with insulin. She delivered locally by semi elective caesarean section at 31+ weeks due to flare up of ulcerative colitis. Postnatally she remained in our DGH with out any complication. Baby's birth weight was 1925 gm with good APGAR.

Discussion

Earlier reports suggests poor pregnancy outcome in patient with CF. Recent data demonstrate good fetal outcome - Live births - 70–90%, Prematurity – 25%; Congenital malformations and miscarriage are similar to general population. No significant difference in survival in women who became pregnant with CF compared to women who did not become pregnant (after adjusting for disease severity)[1].

There is Positive correlation between lung function and maternal outcome. Elective admission of women with poor lung function at 32-34 wks for intensive physiotherapy and possible parenteral antibiotic therapy before anticipated delivery may be required[2].

Our patient was reviewed frequently in the Joint Endocrine Team with close liaison with the tertiary team and gastroenterology team to achieve a successful outcome.

Conclusions

Pregnancy is increasing in CF patients. They should have Preconception counseling. Optimization of maternal health prior to pregnancy can lead to favorable outcome.

Multidisciplinary team management is crucial. In stable cases, pregnancies with CF can be managed in DGH setting with close observation and tertiary support with a low threshold for transfer if required.

References

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