Caring for a Transgender Patient with Cystic Fibrosis

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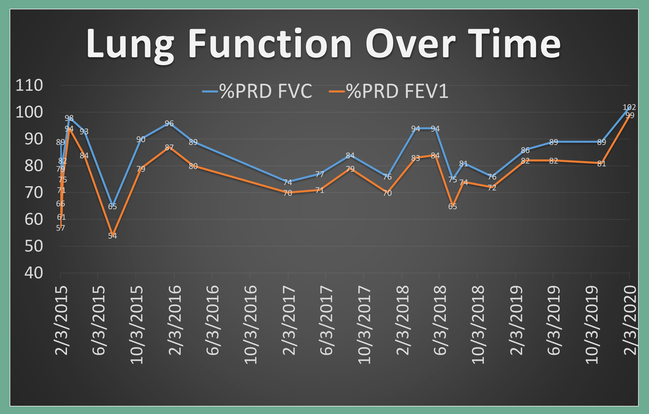
Introduction

- **Transgender individuals are among the patient populations with the least amount of scientific research regarding their care.**
- **Many transgender individuals seek gender-affirming procedures such as Hormone Replacement Therapy (HRT).**
- **There is very little known about the effects of HRT on the respiratory system and even less known about its long-term affects on someone with a chronic respiratory disease such as Cystic Fibrosis.**

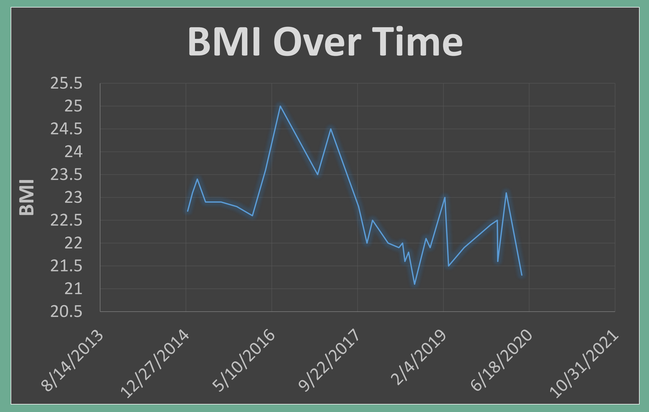
Case Presentation

- **A 23-year-old transgender male was diagnosed from birth with Cystic Fibrosis (delF508 homozygote).**
- **His disease is characterized by obstructive bronchiectasis, cystic fibrosis related diabetes, pancreatic insufficiency, mild cystic fibrosis-related liver disease, osteopenia, attention-deficit-hyperactivity-disorder and depression.**
- **His history is positive for a suicide attempt in June of 2015, cystic fibrosis exacerbations, and *Pseudomonas aeruginosa* and *Staphylococcus aureus* colonization.**
- **The patient disclosed his transgender status to his care team in April 2015 and in June 2016 began his weekly intramuscular injection of 0.5 mL of testosterone cypionate.**
- **In December 2019, patient started elexacaftor-tezacaftor-ivacaftor (Trikafta) and has since shown significant improvement in lung function and respiratory quality of life.**
- **Since onset of HRT, patient has experienced a positive effect in mental health and greater adherence to his medical regimen, but no positive or negative physical effects have been detected.**

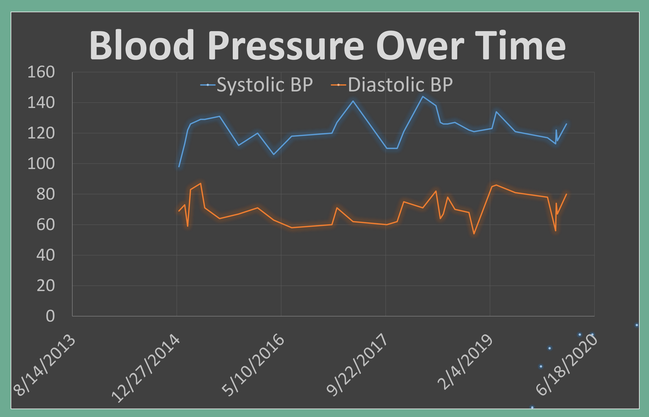
Physical Exam



Graph 1: Percent-predicted FVC and Percent-predicted FEV1 over time



Graph 2: Body Mass Index over time



Graph 3: Systolic and Diastolic Blood Pressure over time

Discussion

- **While it is established Hormone Replacement Therapy increases the risk for certain cardiovascular conditions (such as thromboembolic events and erythrocytosis for estrogen and testosterone supplementation respectively), there is very little existing knowledge about its specific affects on the respiratory system.**
- **Sex hormones in general (particularly estradiol) are shown to affect the respiratory epithelium and therefore warrant further study.**
- **Some studies show that biological women with cystic fibrosis have poorer lung function and worse *Pseudomonas*-related exacerbations than their male counterparts.**
- **Estradiol is thought to be the cause for this "CF gender gap" due to: the increase in infective lung-exacerbations seen when women are at the follicular phase (the highest estradiol producing phase) of their menstrual cycle, the lower rate of lung exacerbations among women taking oral contraceptives, and estradiol's ability to foster mucin production and cause a mutation in the *MucA* gene of *Pseudomonas* thereby increasing its mucoidy.**
- **The handful of testosterone supplementation related studies concerning its affects on the respiratory system at best show a weak association with onset/worsening of obstructive sleep apnea.**
- **Given the lack of studies concerning the long-term effects of hormone replacement therapy on the lung function of someone with cystic fibrosis, healthcare practitioners are without concrete resources to determine the best method of care for patients in this specific situation.**
- **In order to provide the highest quality of care to patients, more studies regarding the long term physical effects of hormone replacement therapy and transgender care in general for cystic fibrosis patients need to be conducted.**

References

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