

Results: Baseline and follow-up OGTTs were available in 65 of 69 participants (33F/32M, median age 18.3, range 12–51.9); 13 were treated with insulin. Percentage HbA1C and fasting glucose improved, but no differences in glucose tolerance were found (Table 1); 17 or 57 participants with abnormal glucose tolerance improved, and nine of 41 without CFRD worsened; hypoglycemia rates and 60- and 120-minute glucose were not different. For the 51 participants for whom complete data were available, no differences in $iAUC_{30}$ glucose, $iAUC_{30}ISR$, $iAUC_{30}ISR:iAUC_{30}$ glucose, or insulin sensitivity were found. These results persisted after adjustment for age, BMI, FEV₁pp, and CFRD history, and although CFRD was associated with lower $iAUC_{30}ISR:iAUC_{30}$ glucose, no interaction between CFRD and visit was found.

Conclusions: In these preliminary analyses, OGTT-related glucose excursion and insulin secretion did not improve with 12 to 18 months of clinically prescribed ELX/TEZ/IVA, despite modest decreases in HbA1C, although HbA1C better reflects longer-term glycemia. The effects of longer ELX/TEZ/IVA treatment on insulin secretion and glucose tolerance remain to be determined.

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Provider education for sexual and reproductive health for adolescents with cystic fibrosis

S. Pope^{1,2}, J. Hamilton^{1,3}, S. Meihls⁴. ¹College of Nursing, University of Utah, Salt Lake City, UT; ²Intermountain Primary Children's Hospital, Salt Lake City, UT; ³Department of Pediatrics, School of Medicine, Salt Lake City, UT; ⁴Pediatric Cystic Fibrosis Clinic, University of Utah, Salt Lake City, UT

Background: Adolescents with cystic fibrosis (CF) are sexually active at the same rate as their healthy peers and have general and disease-specific educational needs regarding sexual and reproductive health. CF care team providers recognize the importance of discussing sexual and reproductive health, but many lack knowledge, comfort, and time to address these topics with adolescents. Providing a clinical practice guideline and education to increase provider confidence in addressing sexual and reproductive health is an important step to implement regular discussions in routine CF clinic visits.

Methods: A quality-improvement project was implemented in the CF clinic in the Intermountain West to increase provider's knowledge and self-efficacy in discussing sexual and reproductive health with adolescents. Provider surveys were reviewed to identify specific educational needs, and previous guidelines were adapted. The clinical practice guideline included information on disease-specific and general sexual and reproductive health topics and a timeline for discussion. The educational module included general tips regarding sexual and reproductive health, mnemonics to assist in discussion, scripting to introduce confidential time during a visit, providing inclusive care, and a clinic-specific referral list. A postintervention survey was sent to care team providers to determine their intent to change practice and to obtain feedback regarding the guideline and education.

Results: Postintervention surveys were sent to nine CF care team members, and the response rate was 22.2% (n = 2). Providers intended to change their practice and implement regular sexual and reproductive health into clinic visits. Providers found the mnemonics and information regarding inclusive care most helpful in increasing their self-efficacy in providing sexual and reproductive health education. Time continues to be the greatest barrier to implementing the guidelines and techniques.

Conclusions: Providers found the guideline and education helpful in increasing their self-efficacy in conversations with adolescents regarding sexual and reproductive health. Further quality improvement work is needed to apply these guidelines to clinical practice and monitor implementation of regular discussion with adolescents in routine visits.

EPIDEMIOLOGY & POPULATION-BASED RESEARCH

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Fetal impact of cystic fibrosis transmembrane conductance regulator modulator use during assisted and natural reproduction and partner pregnancy in men with cystic fibrosis

J. Taylor-Cousar^{1,2}, R. Janney³, T. Kazmerski^{4,5}, D. Tullis⁶, P. Middleton⁷, I. Felton⁸. ¹Division of Pulmonary, Critical Care and Sleep Medicine, National Jewish Health, Denver, CO; ²Division of Pediatric Pulmonary Medicine, National Jewish Health, Denver, CO; ³Clinical Research Services, National Jewish Health, Denver, CO; ⁴Center for Innovative Research on Gender Health Equity, University of Pittsburgh, Pittsburgh, PA; ⁵Department of Pediatrics, University of Pittsburgh Medical Center Children's Hospital of Pittsburgh, Pittsburgh, PA; ⁶Department of Medicine, University of Toronto, Ontario, Canada; ⁷Westmead Clinical School, University of Sydney, Westmead, New South Wales, Australia; ⁸Adult Cystic Fibrosis Department, Royal Brompton Hospital, Guys and St Thomas' NHS Foundation Trust, London, UK

Background: Increasing availability of highly effective cystic fibrosis (CF) transmembrane conductance regulator (CFTR) modulator therapy (HEMT) has improved the quality of life and long-term prognosis for many people with CF. Thus, more people with CF are considering parenthood. Almost all men with CF (MwCF) are infertile because of congenital bilateral absence of the vas deferens (CBAVD). Based on CF animal models, CBAVD occurs early in gestation and is unlikely to be reversible using HEMT, but assisted reproductive techniques (ARTs) can enable MwCF to father children using the sperm in their testes. Animal reproductive models suggest no HEMT teratogenicity, and the amount of exposure of the fetus to HEMT via absorption of seminal fluid through the vaginal wall is predicted to be negligible, although to ensure no sperm exposure to HEMT, the life span of sperm would require MwCF to discontinue CFTR modulators for approximately 3 months before ART. Because abrupt discontinuation of CFTR modulators may result in health decline, MwCF and their providers must consider all potential risks. There are no published data in MwCF regarding use of HEMT during conception and partner pregnancy.

Methods: Beginning in August 2021, CF center staff in the United States, United Kingdom, and Australia completed a two-page anonymous questionnaire regarding MwCF who used CFTR modulators during ART (sperm retrieval and in vitro fertilization) or natural conception with subsequent partner pregnancy.

Results: Providers have submitted 34 surveys for MwCF on CFTR modulators whose partner became pregnant after use of ART (n = 32) or natural conception (n = 2). The median age of the sample was 32 (range 24–43). Fifteen were homozygous for F508del, median percentage predicted forced expiratory volume in 1 second was 76% (range 22–111%), and median body mass index was 24 kg/m² (range 18.5–32.1). Twenty-three were taking elxacaftor/tezacaftor/ivacaftor. The median time that MwCF were taking CFTR modulators before partner conception was 18 months (range 0–82). One newly diagnosed man initiated HEMT after sperm retrieval. Four MwCF stopped CFTR modulators before sperm retrieval, one of whom experienced pulmonary decline. None of the 19 MwCF whose condom use during pregnancy was known used condoms. Fetal complications in partners of MwCF included three first-trimester miscarriages, two* COVID, two breech presentation, two* vaginal bleeding, and one vasa previa. None of the complications were deemed definitively related to use of CFTR modulators. One MwCF experienced testicular infection after sperm retrieval#. Postpartum complications included three# infants with hypoxemia requiring neonatal intensive care unit stay, three maternal blood loss, one forceps delivery, and one caesarean section. No congenital anomalies were reported for any infant. (*/# overlap).

Conclusions: Use of CFTR modulator therapy during partner conception and pregnancy in 34 MwCF has not resulted in higher-than-expected miscarriage rates or congenital anomalies. Providers should consider the risk to the health of MwCF combined with the lack of teratogenicity in animal reproductive models and limited safety data in the human fetus before discontinuing CFTR modulators before ART or natural partner conception. Survey collection is ongoing; results will be updated for presentation.

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