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Intracranial Hypertension in Transgender Patients

Eric Weinlander, MD[1], **Talal Derani, MD**[1], **Wayne T. Cornblath, MD**[1], and **Lindsey B. De Lott, MD, MS**[1]

[1]W K Kellogg Eye Center, Department of Ophthalmology and Visual Sciences, University of Michigan

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Idiopathic intracranial hypertension (IIH) is a potentially blinding condition marked by increased intracranial pressure, papilledema, headaches, and vision loss (1). The role of exogenous sex hormones in the development of IIH is controversial (2). Five cases of IIH occurring in transgender patients undergoing female-to-male (FTM) transition with exogenous testosterone have been reported, suggestive of a causal relationship between exogenous testosterone use and IIH (3). In 3 of the 5 patients, exogenous testosterone was altered or discontinued. We report 2 additional cases in transgender patients: one in a patient undergoing FTM transition and the other in a patient undergoing male to female (MTF) transition. In both, the manifestations of intracranial hypertension were managed entirely without any modification of the patient's hormonal therapies, demonstrating that gender affirming hormonal therapy can be safely continued in transgender patients with IIH.

A 28-year-old obese (body mass index (BMI) 30.13kg/m^2) transgender woman was evaluated in our clinic for optic disc swelling. She had begun oral estradiol (Estrace) and spironolactone for MTF gender transition one year prior, and subsequently developed visual obscurations, frequent headaches and pulsatile tinnitus. Eight months after starting hormone therapy, bilateral optic disc edema was detected and brain MRI and magnetic resonance venography (MRV) showed bilateral transverse sigmoid sinus narrowing. Two lumbar punctures (LP) revealed opening pressures of 30 cm H20 and 39 cm H20 with normal constituents. She was prescribed extended-release acetazolamide 500 mg twice daily, hormone therapy was discontinued, and she was referred to our clinic.

On our examination, visual acuity was 20/15 in each eye without a relative afferent pupillary defect. There was bilateral optic disc edema. Automated visual fields (Humphrey 24-2) showed enlarged blind spots with a mean deviation of −136dB, right eye, and −2.11dB, left eye. On spectral-domain optic coherence tomography (SD-OCT; Spectralis Tracking Laser Tomography, Heidelberg Engineering, Inc.), the retinal nerve fiber layer was thickened bilaterally, measuring 127 μm in each eye.

Correspondence: Lindsey B. De Lott, MD, MS, Kellogg Eye Center, University of Michigan, 1000 Wall Street, Ann Arbor, MI 48105. ldelott@med.umich.edu, Telephone: +1-734-763-9142, Facsimile number: +1- 734-232-2332.

Treatment with acetazolamide was continued. In consultation with the patient and her reproductive endocrinologist, estradiol therapy was resumed. Over the next 5 months, her headaches and papilledema resolved. Her estradiol was ultimately changed by her reproductive endocrinologist to a sublingual formulation in order to further increase her serum estrogen levels and suppress testosterone. She has remained in remission from IIH despite these changes to her hormonal therapy.

A 31-year-old obese (BMI 56.5kg/m^2) transgender man was referred to our clinic for headaches and bilateral optic disc edema. He had previously been taking testosterone injections for FTM gender transition but had been off of testosterone injections for one year prior to presentation. His visual acuity was 20/20 in both eyes, no afferent relative pupillary defect was detected, and Ishihara color plates demonstrated no deficits. Funduscopic examination demonstrated edematous optic discs while automated visual fields (Humphrey 24-2) revealed enlarged blind spots with a mean deviation of −3.80dB, right eye and −3.91dB, left eye. With SD-OCT testing, peripapillary retinal nerve fiber thicknesses measured 385 μm, right eye and 156 μm, left eye. MRI and MRV were unremarkable. LP revealed an opening pressure of 54 cm H2O) with normal constituents. The patient was prescribed extended release acetazolamide 500 mg twice daily. He was then seen by his reproductive endocrinologist, resumed testosterone therapy, and sustained an intentional 20-pound weight loss. Over the ensuing 6 months, his papilledema resolved.

There is no evidence-based consensus guiding the management of hormone therapy in transgender patients with IIH. Given the poorly-understood, and possibly causative role of sex hormones in the development of IIH, clinicians may be inclined to alter gender-affirming hormone therapy once the diagnosis of IIH is made. Of the five previously reported cases of IIH developing in FTM patients receiving testosterone, the dose testosterone was reduced in 2 patients and changed to an alternate formulation in a third (3). Similarly, in our patient undergoing MTF transition, estradiol was stopped prior to our evaluation.

Remission from IIH was achieved in both of our patients while continuing hormone therapy. In the patient undergoing MTF transition, hormone therapy was safely resumed and subsequently increased to suppress endogenous testosterone; she has remained in remission on acetazolamide. Likewise, in the patient undergoing FTM transition, IIH resolved with weight loss and acetazolamide, and he has remained in remission even after resumption of regular testosterone injections.

Although it is uncertain if hormone therapy can precipitate IIH, we report two instances of IIH in transgender individuals that were successfully treated without alteration to the patient’s gender-affirming hormone therapy. Patients with IIH undergoing hormone therapy for gender transition may still be responsive to conventional treatment without the need for change in hormone therapy.

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