

Transvestism recognized in Ehlers-Danlos syndrome: Report of two cases



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With dual certification as clinical geneticists and psychiatrists, we have the special opportunity to treat patients with Ehlers-Danlos syndrome (EDS) complicated with mental disorders in Japan. EDS comprises a series of rare hereditary connective tissue diseases characterized by musculoskeletal, skin, and cardiovascular involvements. EDS may be associated with physical as well as psychological sufferings that can lead to psychiatric problems. EDS imposes substantial psychological burden on patients, and recent large-scale studies have suggested that patients with EDS have a higher risk of mood disorders than the general population. We report on two Japanese patients with EDS complicated with mood disorders; they secondarily developed transvestism, also termed cross-dressing, that was rooted to early stressful situations from childhood through adolescence. This report is the first on EDS with transvestism to the best of our knowledge, which might be encountered by public health professionals. As EDS involves uncomfortable or even distressing symptoms, the two patients could have been experiencing incongruent feelings in personality as well as gender identity. Physicians are advised to be aware of various potential psychological and psychiatric issues that may accompany EDS. Informed consent was obtained from both of the patients with assurances of preserving anonymity and privacy.

Keywords: transvestism, mood disorder, gender dysphoria

Classification for EDS subtypes

- ★ **Mostly, normal intellectual development**
- **Classical Classification:** more patients with EDS in IX type
- **Present Classification(2017):** No specific types regarding intellectual developmental disorder or psychiatric symptoms

EDS and mental disorders/ symptoms in the references;

- ✓ 42.5% patients with EDS have psychiatric disorders such as anxiety disorder and major depressive disorder. (Hershenfeld, AM J Psychiatry 2016)
- ✓ Risks for developmental disorders and mood disorders are higher in EDS patients than in general population. (Cederlof, BMC Psychiatry 2016)
- ✓ Depressiveness induced by chronic pain and physical disability are often seen in Hypermobility type EDS. (Baeza-Velasco, Disability and Rehabilitation 2019)
- ✓ Trait anxiety scores did significantly correlate with both state anxiety and hypermobility scores. (Mallorqui-Bague, Eur Psychiatry 2015)
- ✓ psychotropic agents users: 41.4% of EDS pat. vs. 13.9% of control. (Bulbena, Gen Hosp Psychiatry, 2011)

Other psychiatric phenotypes more seen in EDS patients;

- ✓ Substance user (Alcohol, Tobacco, analgesics) → anxiety relief (Baeza-Velasco, Rheumatol Int. 2015)
- ✓ Eating disorder (Bulbena, Am J Med Genetics 2017)

Few cases have been reported regarding schizophrenia with EDS.

Terminology for Transvestism: not a mental health condition

- recurrent, intense sexual arousal from cross-dressing
- an individual's wearing of clothes that are typically associated with a different gender from the one with which that person identifies
- the individuals are wrongly assumed that their transvestism are homosexual
- a majority of individuals who identify as transvestites are heterosexual men who dress in women's clothing to express an inner femininity or desire to be seen as a beautiful

≠ Transvestic disorder, ≠ Gender dysphoria (DSM-5 criteria)

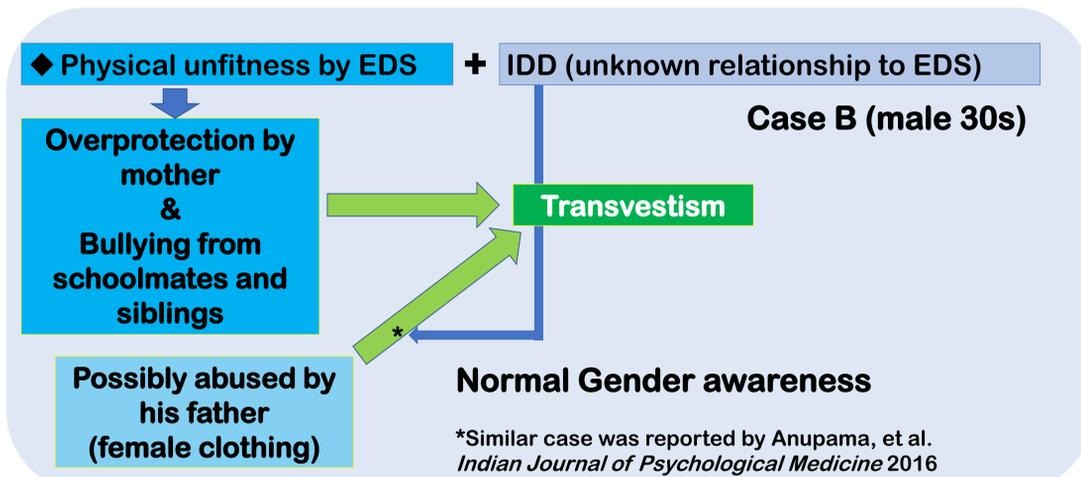
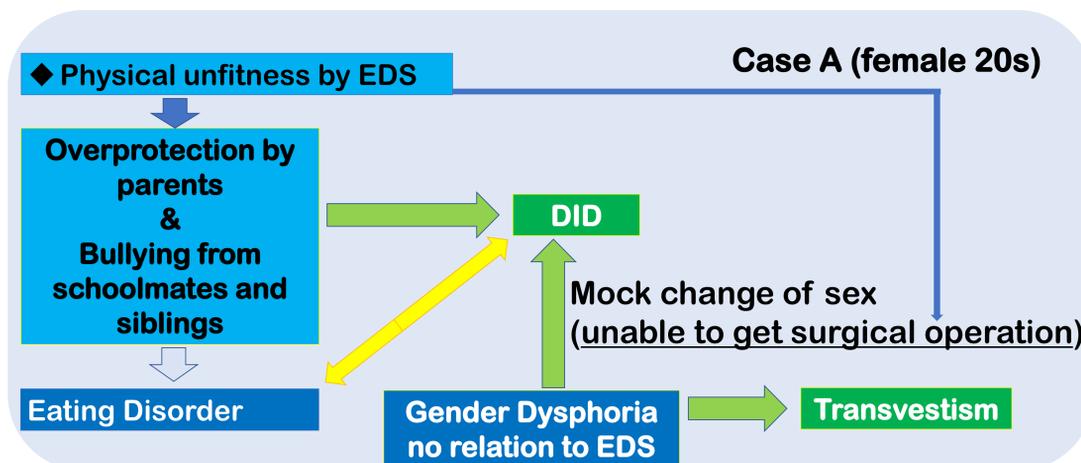
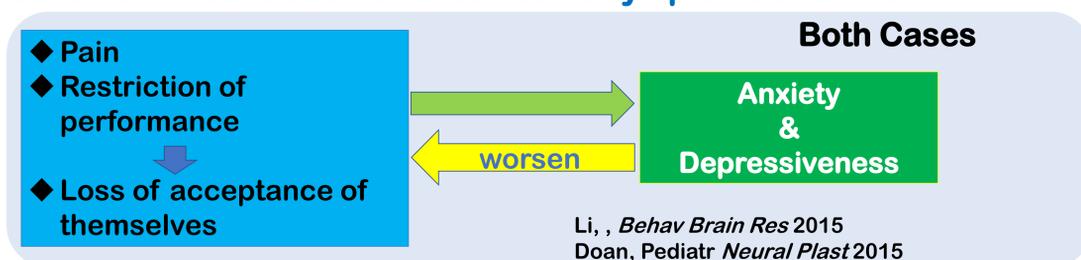
2 cases and EDS criterion (ref. AJMG 175C : 8-26,2017))

Classical EDS	Case A	Case B
Autosomal Recessive	Concordant monozygotic twin	sporadic
• Major criteria		
1. Skin hyperextensibility and atrophic scarring	+	+
2. Generalized joint hypermobility (GJH)	+	+
• Minor criteria		
1. Easy bruising	+	-
2. Soft, doughy skin	+	+
3. Skin fragility (or traumatic splitting)	+	+
4. Molluscoid pseudotumors	-	-
5. Subcutaneous spheroids	-	-
6. Hernia (or history there of)	-	-
7. Epicanthal folds	+	-
8. Complications of joint hypermobility	+	+
9. Family history of a first degree relative	+	-
Psychiatric symptoms	Gender dysphoria DID Eating disorder	Transvestism Bipolar Disorder IDD (IQ=66)

*IDD: Intellectual developmental disorder

*DID: Dissociative identity disorder

Schema of association between some symptoms and EDS



Conclusion

Psychological issues and mood disorders often complicate the course of patients with EDS. The association with the negative aspects of EDS affects the patients' social lives and dimensional identification issues. Based on the multidimensional nature of quality of life, medical professionals need to pay special attention to patients with EDS regarding evaluation and treatment. It is necessary to consider the patient's social and psychological stress based on individual background for the purpose of EDS genetic counseling, as well as for medical management of psychiatric symptoms of EDS.