

A RARE CASE OF A 46,XX, SRY-NEGATIVE MALE WITH SPERM PRODUCTION

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Objective: To report the first case of 46,XX,SRY negative male with sperm production.

Design: A case report.

Setting: Reproductive medicine department of a women's & children's hospital.

Patient: A 33 year old male found to have 46,XX disorder of sex development(DSD) during investigation of secondary subfertility.

Results: A couple with a previous spontaneous pregnancy presented with secondary subfertility. The male was diagnosed to be 46,XX, SRY-negative on 2 independent karyotype analysis. He was well masculinized and had sperms on semen analysis.

Conclusions: To the best of our knowledge, this is the first reported case of 46,XX, SRY-negative male with sperm production and a spontaneous pregnancy.