

Gonadectomy for Adults With DSD Conditions In The International Disorders of Sex Development Registry

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Introduction

Depending on the underlying diagnosis, Disorders of Sex Development (DSD) can be associated with an increased risk of germ cell cancers. To date, however, knowledge regarding the indications and timing of gonadectomy is lacking. The International Disorders of Sex Development (I-DSD) Registry offers the opportunity to investigate these outcomes, with the overall aim to improve clinical care for affected individuals.

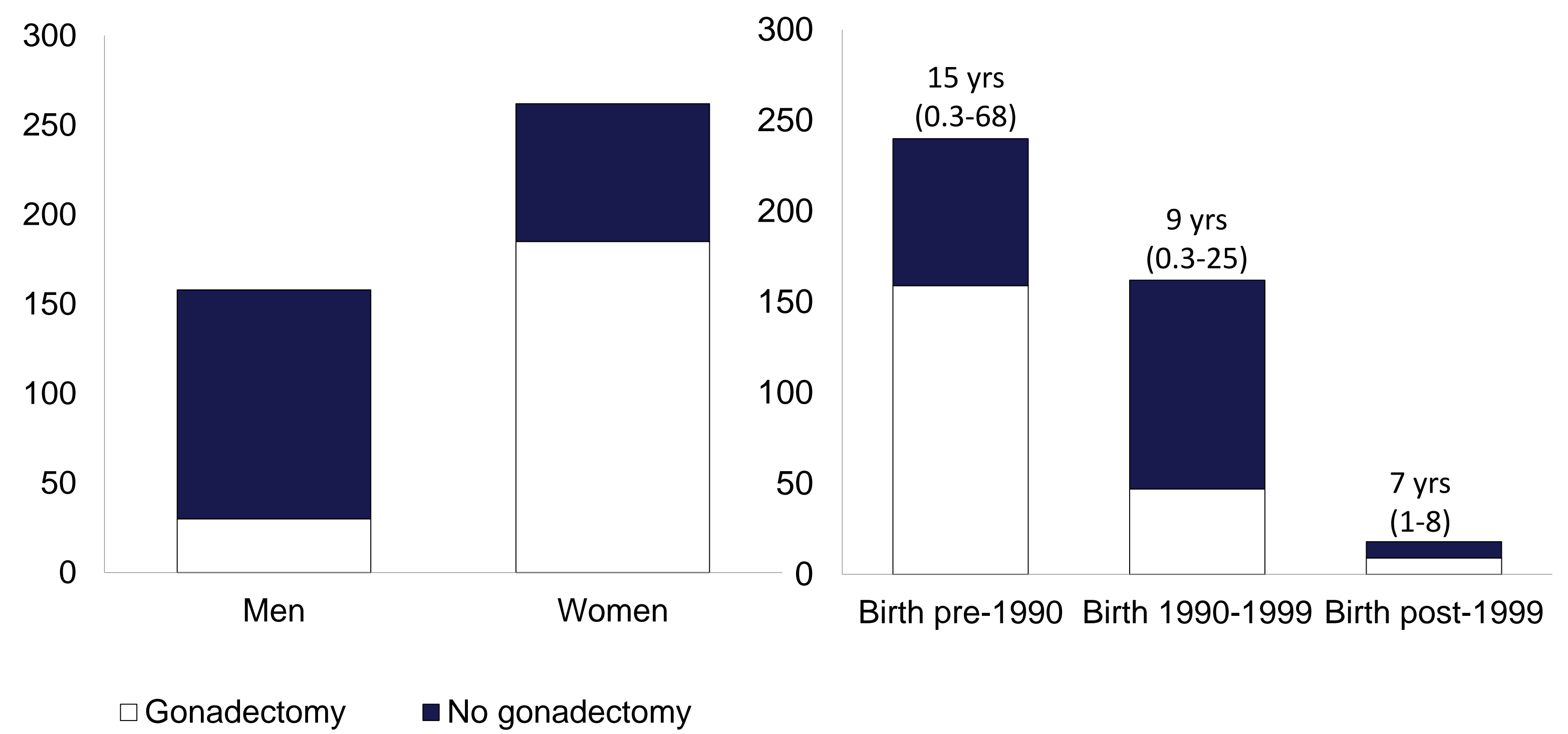
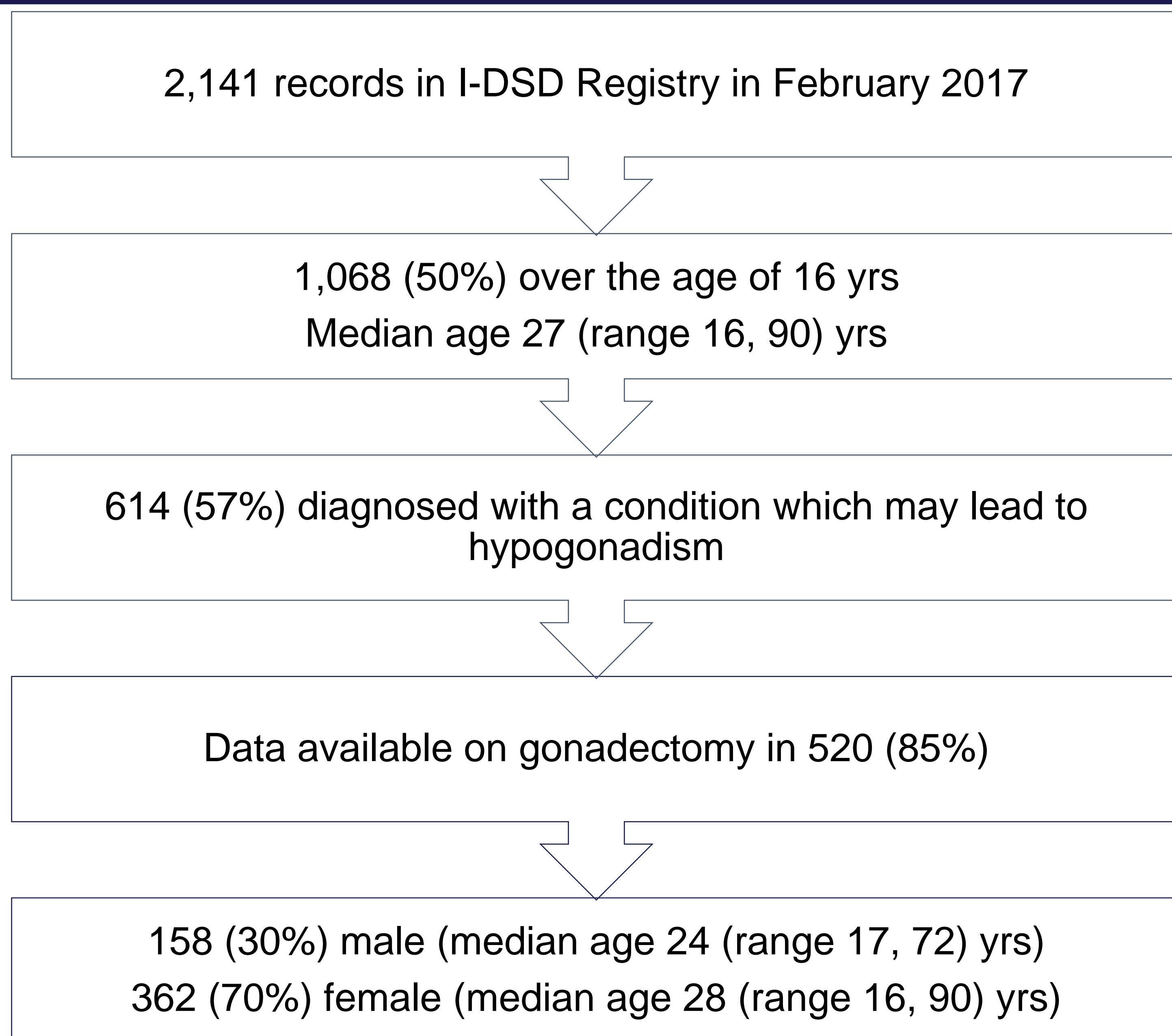
Aim: To investigate the prevalence of gonadectomy in individuals with conditions which could lead to hypogonadism on the I-DSD Registry.

Methods

The I-DSD Registry and its users were approached to identify all participants aged 16 years or over at the time of data collection who had one of the following diagnoses which could lead to hypogonadism:

- Disorder of androgen action
- Disorder of androgen synthesis
 - Gonadal dysgenesis
- Non-specific disorder of undermasculinisation (NSUMD).

Results



Condition	Females with gonadectomy (%)	Median age female gonadectomy (range) (years)	Males with gonadectomy (%)	Median age male gonadectomy (range) (years)
Complete androgen insensitivity syndrome (CAIS)	123/154 (79.8)	15 (0.3-68)	0/0 (0)	
Complete gonadal dysgenesis (CGD)	55/69 (79.7)	15 (0.3-21)	2/7 (28.6)	5 (4-5)
Non specific disorder of undermasculinisation (NSDUM)	6/6 (100)	14 (3-26)	3/22 (13.6)	9 (6-10)
Partial androgen insensitivity syndrome (PAIS)	26/29 (89.6)	12 (1-24)	3/41 (7.3)	32 (10-54)
Partial gonadal dysgenesis (PGD)	23/26 (88.4)	2 (0.3-21)	15/51 (29.4)	1 (0.1-13)
17β hydroxyteroid dehydrogenase deficiency (17BHSD)	25/25 (100)	11 (0.5-21)	0/1 (0)	
5α reductase deficiency (5ARD)	11/14 (78.6)	6 (2-17)	0/5 (0)	
Other	16/39 (41)	16 (1-21)	7/31 (22.6)	17 (10-26)

- Females had gonadectomy at a later age (median 14 (range 0.3, 68) years) compared to males (median 5 (range 0.1, 54) years) (p=0.047).
- Gonadectomy was performed later in males (median 15 vs 4 years, p=0.0004) and females (median 17 vs 8 years, p<0.0001) after the publication of the 2006 consensus statement on the management of DSD conditions.

Summary and conclusions

- Rates of gonadectomy vary from one diagnosis to another.
- Gonadectomy is performed at a later stage since the publication of the 2006 consensus statement on the management of DSD conditions.
- A substantial proportion of young men and women with a range of DSD continue to retain gonads into adulthood.