Title: How is Psychosocial Adaptation and Cognitive Function Assessed in Differences/Disorders of Sex Development?

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Introduction:
Differences/disorders of sex development (DSD) are defined as “congenital conditions in which development of chromosomal, gonadal, or anatomic sex is atypical” [1]. As such, they comprise a wide range of discrete medical conditions (see Table 1) and have a prevalence of approximately 1 in 5000 live births [2]. Some DSD, such as Klinefelter syndrome [3-16] and Turner syndrome [17-38], are known to be associated with characteristic psychological profiles associated with the genotype of their condition. While others, such as girls and women with congenital adrenal hyperplasia (CAH), may experience cognitive difficulties due to prophylactic interventions to prevent genital virilization [39]. Studies examining most other diagnoses that fall under the umbrella of DSD cover an extremely broad range of psychosocial and cognitive concerns.

Table 1. Example of a DSD Classification System.

Sex Chromosome DSD	46,XY DSD	46,XX DSD
A: 47,XXY (Klinefelter syndrome) and variants B: 45,X (Turner syndrome) and variants C: 45,X/46,XY (mosaicism) and variants D: 46,XX/46,XY (chimerism)	A: Disorders of gonadal (testis) development Complete or partial gonadal dysgenesis (e.g., *SF1/NR5A1, WT1, GATA4, FOG2/ZFPM2, CBX2, SRY, SOX9, SOX8, MAP3K1, ESR2/NR3A2, DMRT1, TSPYL1, DHH, SAMD9, ARX, MAMLD1/CXorf6*) Ovotesticular DSD Testis regression	A: Disorders of gonadal (ovary) development Gonadal dysgenesis Ovotesticular DSD (e.g. *NR5A1, NR2F2, RSPO1*) Testicular DSD (e.g., *SRY+*, dup *SOX9*, dup *SOX3, NR5A1, NR2F2, RSPO1, WNT4*)
	B: Disorders in androgen synthesis or action Disorders of androgen synthesis Luteinizing hormone (LH) receptor mutations Smith-Lemli-Opitz syndrome StAR protein mutations Cholesterol side-chain cleavage *(CYP11A1)* 3β-hydroxysteroid dehydrogenase 2 *(HSD3B2)* 17α-hydroxylase/17,20-lyase *(CYP17)* P450 oxidoreductase *(POR)* Cytochrome b_5 *(CYB5A)* Aldo-keto reductase 1C2 *(AKR1C2)* 17β-hydroxysteroid dehydrogenase *(HSD17B3)* 5α-reductase 2 *(SRD5A2)* Disorders of androgen action Androgen insensitivity syndrome Drugs and environmental modulators	B: Androgen excess Fetal 3β-hydroxysteroid dehydrogenase 2 *(HSD3B2)* 21-hydroxylase *(CYP21A2)* P450 oxidoreductase *(POR)* 11β-hydroxylase *(CYP11B1)* Glucocorticoid receptor mutations Fetoplacental Aromatase (CYP19) deficiency Oxidoreductase (POR) deficiency Maternal Maternal virilizing tumors (e.g., luteomas) Androgenic drugs
	C: Other Syndromic associations of male genital development (e.g., cloacal anomalies, Robinow, Aarskog, hand-foot-genital, popliteal pterygium) Persistent müllerian duct syndrome Vanishing testis syndrome Isolated hypospadias Cryptorchidism *(INSL3, GREAT)* Environmental influences	C: Other Syndromic associations (e.g., cloacal anomalies) Müllerian agenesis/hypoplasia (e.g., MKRH) Uterine abnormalities (e.g., MODY5) Vaginal atresias (e.g., McKusick-Kaufman) Labial adhesions

CYP, Cytochrome P450 isoenzyme; *DSD*, disorders of sex development; *MODY5*, maturity-onset diabetes of the young type 5; *MRKH*, Mayer-Rokitansky-Küster-Hauser; *StAR*, steroidogenic acute regulatory (protein).

Note. Table from Chan, Y.-M., Hannema, S. E., Achermann, J. C., & Hughes, I. A. (2020). Disorders of Sex Development. In S. Melmed, R. J. Auchus, A. Goldfine, B., R. J. Koenig, & C. J. Rosen (Eds.), Williams Textbook of Endocrinology (Fourteenth ed., pp. 867-936.e814). Elsevier

In addition to evaluating individuals with a DSD, the psychosocial and cognitive adaptation of their parents is also clinically relevant. Research examining parental functioning has scrutinized numerous psychosocial constructs, including stress, anxiety, depression, social support, quality of life, and illness uncertainty [40-42]. However, as in the DSD patient population, assessment methods differ by study.

For the purposes of this scoping review, psychosocial adaptation is being defined broadly to cover constructs related to both psychological functioning (e.g., anxiety, depression, health-related quality of life) and social functioning (e.g., romantic relationships, quality of life). Cognitive adaptation is being defined as the ability to perceive, learn, and recall events and information.

An improved understanding of the psychosocial and cognitive adaptation of patients with a DSD and their parents requires the ability to compare findings across studies. However, these comparisons are either facilitated or hampered by the comparability of measures used across studies. Unfortunately, psychosocial assessment of patients with a DSD is inconsistent and further complicated by the international nature of the extant literature and that it spans across multiple disciplines. The breadth of data available make it difficult to distill down the overarching psychosocial needs of this population. Despite this barrier, psychologist are tasked with supporting this population as part of multi/interdisciplinary teams of specialty providers [1].

Although there are meta-analyses or systematic reviews examining psychosocial functioning among subsets of DSD diagnoses/cohorts (see [17 28 43 44]), the focus of these projects remain narrow in condition/group examined (e.g., only Turner syndrome, only those with 46,XY DSD, or only patients who had undergone surgical intervention for their DSD). A preliminary search for scoping and systematic reviews on psychosocial functioning across DSD was conducted on 2020 and again in 2021 with no studies identified. Databases searched included JBI Database of Systematic Reviews and Implementation Reports, Cochrane Database of Systematic Reviews, Cumulative Index to Nursing and Allied Health Literature (CINAHL), PubMed, and PsycINFO through the University of Michigan library system.

The aim of this scoping review is to ascertain the methods used to assess psychological and cognitive variables and outcomes across the range of DSD for both patients and their parents. It is hypothesized that there will be a broad range of constructs and measures utilized, with little consensus across studies/sites.

Inclusion Criteria:
Quantitative and qualitative studies published after the publication of the Consensus Statement [1] will be included in the scoping review if they examine the psychosocial adaptation and/or cognitive function of children, teenagers, and adults with a DSD and/or their parents/caregiver. Quantitative and qualitative data will be included. Exclusion criteria will include first person accounts of psychosocial adaptation and/or cognitive function and any articles not written in English.

Types of participants:

- Children, teenagers, and adults with a DSD as defined by the Consensus Statement [1].
- Parents/caregivers of an individual with a DSD as defined above.
- Non-affected adults (e.g., healthcare providers, teachers, romantic partners of an individual with a DSD) or children (e.g., siblings of an individual with a DSD), as long as they are reporting on the psychosocial/cognitive functioning of the individual with DSD or the individual with DSD's parent/caregiver.

Concept:
The concept of interest for this scoping review is understanding the assessment measures and methods used to evaluate psychosocial and cognitive functioning in DSD.

Context:
No particular context will be applied to this project. However, sources will be limited to those written in English. As such, this scoping review will include studies published from any cultural or regional setting that is written in the English language.

Types of evidence sources:
We will include the following study designs:

Quantitative and qualitative primary research studies will be included in the scoping review. This will include randomized controlled trials, time series analyses, non-randomized studies, and observational studies, including controlled before-after studies, and pre-post studies.

The following will be excluded: literature reviews, commentaries, book chapters, and editorials.

Methods:
The methods used in this scoping review will follow the frameworks proposed by Arksey and O'Malley [45] and Levac and colleagues [46], using the methods outlined in the JBI Manual for Evidence Synthesis [47]. The review team followed a multi-step, iterative process for developing and refining the search strategy.

Search Strategy:

Description of strategy:

The review team met with two informationists (LJ, KS) in early 2020. Using a short list of sentinel articles provided by the review team, the informationists were able to craft an initial search strategy that was used to inform the selection of potential databases, concepts, and search terms. The search strategies were developed to identify published primary studies. The databases that were selected for this project include Cochrane Library, PubMed, Ovid MEDLINE (Ovid MEDLINE(R) and Epub Ahead of Print, In-process & Other Non-INdexed Citations, Daily and Versions(R)), (ELSEVIER) Embase, (EBSCO) CINAHL Complete, (EBSCO) PsycInfo, (EBSCO) LGBT Life, and (ELSEVIER) Scopus.

As a group, the team reviewed preliminary searches for scoping and systematic reviews on psychosocial functioning across DSD in Cochrane Library and PubMed. This initial search was also used to identify relevant concepts, controlled vocabulary, and keywords.

After initial search strategies were analyzed and refined, it was later determined that Ovid MEDLINE would be the preferred database for searching MEDLINE. As the searches were translated across the remaining databases, the entire team reviewed search terms and results for each database, and provided feedback on controlled vocabulary and keywords. The final search strategy was built around three main concepts: disorders of sex development, psychological and social functioning, and assessment. Due to the publication of the Consensus Statement [1] in 2006, the review team was only interested in articles published after 2006, so the publication date limit was applied across all databases. The review team was also unable to provide translation for articles in languages other than English, so an English language limit was also applied to the searches. When available, publication limits were applied to exclude reviews, commentaries, and book chapters. The

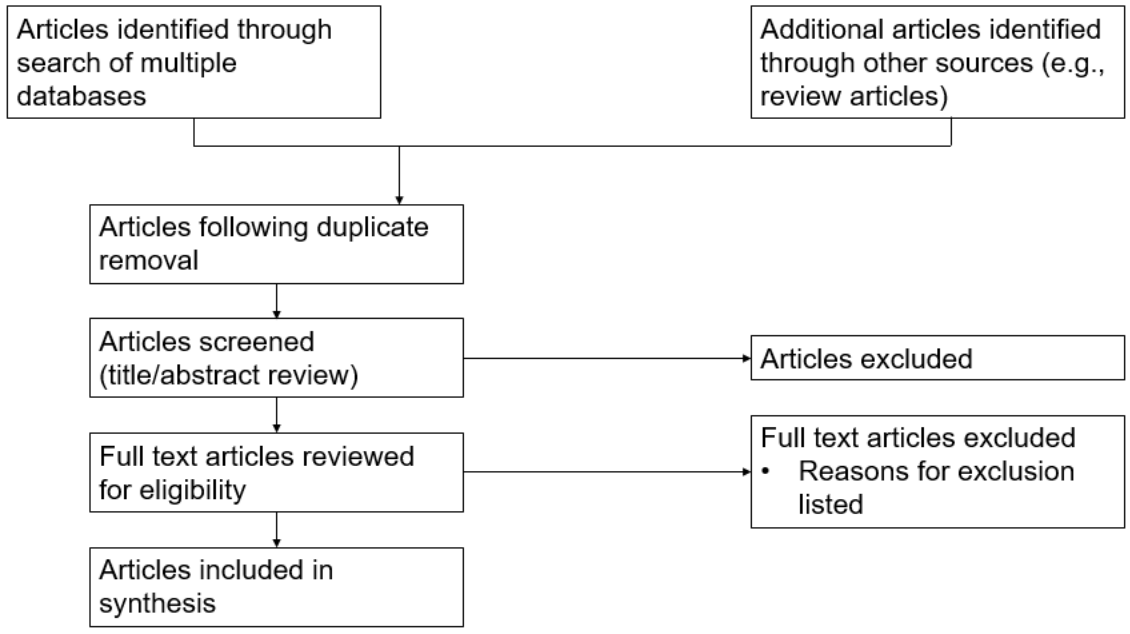
final searches were run on 12/3/2020 through 12/7/2020, and EndNote X9 was used to manage citations, and to identify and remove duplicates.

A complete search strategy for Ovid MEDLINE has been included in Appendix A.

Supplemental strategies:
A hand-search for non-indexed and difficult to locate studies will be conducted, including examining key journals. We will also scan the reference lists of all included articles.

Source of evidence selection:
The review of sources will utilize the program DistillerSR. Article selection will be based on the inclusion/exclusion criteria described above and will include a review of title and abstract, followed by a full-article review. All reviewers will undergo a training process including reading relevant articles, for example [48-50], and looking over the codebook developed for this project. Pilot testing will take place, including the entire review team completing a title/abstract and full-text review of 25 randomly selected articles, with the use of the inclusion/exclusion criteria and the codebook. Throughout pilot testing, the review team will meet to review and update inclusion/exclusion criteria and codebook. Screening will commence once all 25 articles have been reviewed and discussed and when there is at least 75% agreement among reviewers. When completing the screening, at least 2 reviewers will review each source at each level (title abstract and full-article review) and disagreements will be reconciled by consensus or by a third reviewer. See Figure 1 for flowchart of review process.

Figure 1. Review process



Details will be provided in a table for all articles identified for inclusion in the final synthesis for this scoping review. Reasons for exclusion will also be provided about excluded articles.

Data extraction:
The data extraction form will extract the following key information from each article:
1. Author(s)

2. Year of publication
3. Country of origin (where each study was conducted)
4. Aims/purpose
5. Population and sample size
    a. DSD diagnosis/identity, race, ethnicity, sexual orientation
6. Methodology / methods
7. Psychosocial / cognitive outcomes and details of how these were measured

The data extraction form will be utilized during the pilot phase of the project and further refined, as needed. If additional data is determined to be needed during the screening and data extraction process, the data extraction form will also be updated.

Analysis of the evidence:
This scoping review will be analyzed using frequency data.

Presentation of the results:
Results of the scoping review will be presented in the format provided in Table 2.

Table 2: Example tabular presentation of data

Parameter	**Results**
Numbers of publications	Total number of sources of evidence Total numbers between 2006 and December 2020 Number of publications every year
Types of studies	Randomized controlled trials Non-randomized controlled trials Quasi-experimental studies Before-and-after studies Prospective cohort studies Retrospective cohort studies Case-control studies Cross-sectional studies Other quantitative studies Qualitative studies Mixed methods studies
Population(s) identified	Age: • Children 0-17 years • Adults >18 years • Parent/s and/or caregivers of a child with a DSD DSD Diagnoses/Identity
Psychosocial / Cognitive domains	Psychosocial: • Depression, anxiety, quality of life, gender identity, etc Cognitive: • Attention, memory, processing speed, adaptive functioning, academic functioning, IQ, etc

Assessment type	Standardized questionnaire Non-standardized items/questionnaire Qualitative interviews Other methodology
Assessment measure	Measure(s)/assessment battery utilized

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Appendix A

Ovid MEDLINE

Ovid MEDLINE(R) and Epub Ahead of Print, In-process & Other Non-INdexed Citations, Daily and Versions(R)

1.
(17-Hydroxysteroid Dehydrogenase Deficiency OR Anorchia).rs. OR exp Cloaca/ OR exp Disorders of Sex Development/ OR exp Hypospadias/ OR exp Intersex Persons/ OR exp Kallmann Syndrome/ OR exp Klinefelter Syndrome/ OR exp Ovotesticular Disorders of Sex Development/ OR exp Turner Syndrome/ OR exp WAGR Syndrome/ OR Mullerian aplasia.rs. OR Mullerian Ducts/ab OR Penis agenesis.rs. OR (17 beta hydroxysteroid dehydrogenase OR 21-hydroxylase deficiency OR 5 alpha reductase deficiency OR 5-alpha-reductase-2 OR 5alpha-rd2 deficiency OR 5rd2 deficiency OR adrenal hyperplasia OR adrenogenital syndrome OR androgen insensitivity syndrome OR anorchia OR aphallia OR clitoromegaly OR cloaca OR cloacal exstrophy OR disorders in androgen synthesis OR empty scrotum OR gonadal regression OR hermaphrodite OR hermaphrodites OR hermaphroditism OR hermaphroditismus OR hypospadia OR hypospadias OR intersex OR intersexualities OR intersexuality OR kallmann's syndrome OR kallmanns syndrome OR luteinizing hormone receptor mutation OR mayer rokitansky kuster hauser syndrome OR macroclitoris OR micro-penis OR micropenis OR microphallus OR ovotestes OR ovotesticular OR pseudohermaphrodite OR pseudohermaphrodites OR pseudohermaphroditism OR sex chromosome mosaicism OR sex reversal OR swyer syndrome OR testicular feminization syndrome OR turner's syndrome OR turners syndrome OR vanishing testes OR wagr OR wagro).mp. OR ((sex OR sexual) adj3 (difference OR differences OR differentiation) adj3 (development OR developments)).mp. OR ((sex OR sexual) adj3 (difference OR differences OR differentiation) adj3 (disorder OR disorders)).mp. OR ((sex OR sexual) adj3 (development OR developments) adj3 (disorder OR disorders)).mp. OR ((atypical OR atypia OR ambiguous OR ambiguity OR ambiguities) adj3 (genitalia OR genital OR genitals)).mp. OR (((penis OR penile OR clitoris OR mullerian OR gonadal OR uteri OR uterus OR uteruses OR uterovaginal OR vagina OR vaginal OR testis OR testes OR testicular) adj3 (dysgenesis OR agenesis OR atresia OR aplasia OR hypoplasia OR regression OR absence OR absent OR vanishing)) OR ((uteri OR uterus OR uteruses) adj3 (didelphys OR bicornus OR absent))).mp.

2.
exp Gender Identity/ OR exp Social Integration/ OR exp Cognitive Science/ OR exp emotional intelligence/ OR exp Emotions/ OR exp Interpersonal Relations/ OR exp Mental Disorders/ OR exp Neuropsychiatry/ OR exp neuropsychology/ OR exp Sexuality/ OR Sexual Behavior/ OR (adjustment disorder* OR affect disorder OR agoraphobia OR alcohol-related disorder* OR anger OR anorexia nervosa OR antisocial personality disorder OR anxiety OR anxiety disorder* OR apathy OR attention deficit disorder* OR behavior disorders OR binge-eating disorder* OR bipolar disorder* OR body dysmorphic disorder* OR body integrity identity disorder* OR bulimia nervosa OR castration anxiety OR child behavior disorder* OR cognition disorder* OR cognition OR cognitive science* OR communication disorder* OR community integration* OR

conversion disorder OR cyclothymic disorder OR dependent personality disorder OR depression OR depressive disorder OR developmental disabilities OR disgust OR disruptive disorders OR impulse control disorders OR conduct disorders OR dyspareunia OR dyssomnias OR elimination disorders OR emotional adjustment OR emotional intelligence* OR emotional regulation OR empathy OR encopresis OR enuresis OR erectile dysfunction OR fear OR eating disorders OR firesetting behavior OR gender dysphoria OR gender issue OR gender relation OR gender role OR intellectual disability OR interpersonal relation OR irritable mood OR learning disabilit* OR mental disorder OR mood disorders OR motivation OR motor disorders OR motor skills disorders OR mutism OR neurocognitive disorders OR neurodevelopmental disorders OR neuropsychiatry OR neuropsycholog* OR neurotic disorders OR neurotic disorders OR obsessive-compulsive disorder OR panic OR paraphilic disorders OR parasomnias OR pedophilia OR performance anxiety OR personality disorders OR pervasive child development disorders OR phobic disorders OR premature ejaculation OR (psychiatric adj5 (disease* OR illness*)) OR psychiatric diagnosis OR psychiatric diagnosis OR psychiatric disease OR psychiatric disorder* OR psychiatric fetishism OR psychiatric illness* OR psychological distress OR psychological sexual dysfunction* OR psychosocial OR sadness OR separation anxiety OR severe mental disorder* OR sexual activity OR sexual disorder* OR gender disorder* OR sexual behavior OR sexual functioning OR sexual orientation OR shame OR social intelligence* OR social interaction* OR somatoform disorder* OR tic disorder* OR transvestism OR trauma disorder* OR stress disorder* OR vaginismus OR voyeurism).mp.

3.
exp Diagnostic Self Evaluation/ OR exp Psychiatric Status Rating Scales/ OR exp Qualitative Research/ OR exp "Surveys and Questionnaires"/ OR exp Interviews as Topic/ OR exp Interview, Psychological/ OR exp Personality Inventory/ OR exp self report/ OR
(quantitative or qualitative or scale or scales or questionnaire or questionnaires or survey or surveys or surveyed or interviews or interviewed or interview or measure or measured or measures or measurement or assessment or score or scores or scored or scoring or mixed method or ((self or patient or patients) adj2 (report or reported or reports or reporting))).mp.

4.
(animals.sh. NOT humans.sh.)

((1 AND 2 AND 3) NOT 4) 1457 results 12/3/20
Limits:
English
2006 - current
917 results 12/3/2020