

Subjective need for psychological support (PsySupp) in parents of children and adolescents with disorders of sex development (dsd)

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Background:

Disorders/diversity of sex development (dsd) is an umbrella term for rare congenital conditions with incongruence between chromosomal, gonadal and phenotypic sex often diagnosed within childhood. As most parents are unprepared for this complex and challenging situation psychological support (PsySupp) is recommended.

The aim of this study was to analyse the subjective need for PsySupp in parents of children and adolescents with dsd and to identify associated variables.

Subjects and Methods:

Evaluation of the need for PsySupp in parents of 329 children with dsd from the German clinical evaluation study of the German DSD network sponsored by BMBF from 2005-2008. A self-constructed "dsd-questionnaire" was used. Exploratory data analysis was undertaken.

| Diagnoses of children and adolescents were classified into dsd-46-XX or 46-XY without (c) or with partial (p) androgen effects, and female (f) or male (m) sex of rearing. | |
|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| dsd-XX-p-f n = 132 | CAH (128), partial/mixed gonadal dysgenesis (1), complex malformation (1), aromatase deficiency (2) |
| dsd-XY-p-f n = 66 | pAIS (12), partial/mixed gonadal dysgenesis (29), 17beta-hydroxysteroid- dehydrogenase III deficiency (7), 5 alpha-reductase II deficiency (4), LH-receptor defect (1), 17/20-Lyase deficiency (1); severe hypospadias (1); complex malformation (5), penis agenesis (1), cloacal extrophy (1), unclassified clinical diagnoses of dsd (4) |
| dsd-XY-p-m n = 108 | severe hypospadias (50), pAIS (9); partial/mixed gonadal dysgenesis (21); 5alpha-reductase II deficiency (3), LH-receptor defect (3), 17beta hydroxysteroid-dehydrogenase III deficiency (2), not precisely classified (2), disorder of anti-müller hormone (1), Complex malformation (3), 46, XX-male*(3), micropenis (2), unclassified clinical diagnoses of dsd (9) |
| dsd-XY-c-f n = 22 | cAIS (12), complete gonadal dysgenesis (9), clinically most likely SF1 mutation, but defect was not found (1) |

AIS= androgen insensitivity syndrome; CAH= congenital adrenal hyperplasia, cAIS= complete androgen insensitivity syndrome, dsd= disorders/diversity of sexual development, pAIS= partial androgen insensitivity syndrome, + incl. mosaics with parts of „Y chromosome“ (in some cases chromosome status has not been investigated, in these cases classification results according the clinical status), *Included in XY-dsd-p-m despite karyotype 46,XX; Excluded from diagnosis group comparisons: one child with 46, XX & without androgen effects

Table 1 Diagnoses of the children and adolescents

| | How did you assess your child's genitalia immediately after birth?* | | | | | |
|------------|---------------------------------------------------------------------|------------------------|-----------|------------------------|--------|------------------------|
| | male | | ambiguous | | female | |
| | n | Need for PsySupp n (%) | n | Need for PsySupp n (%) | n | Need for PsySupp n (%) |
| dsd-XX-p-f | 30 | 13 (43.3) | 34 | 9 (26.5) | 57 | 16 (28.0) |
| dsd-XY-p-f | 8 | 5 (62.5) | 14 | 8 (57.1) | 40 | 24 (60.0) |
| dsd-XY-p-m | 72 | 24 (33.3) | 19 | 8 (42.1) | 9 | 9 (100.0) |
| dsd-XY-c-f | 0 | | 0 | | 21 | 8 (36.4) |

Not significant

Table 3: Parents' perception of their child's genitalia & need for PsySupp

| Total of parents | 317 | % |
|-----------------------------------------------------------------------------------|------------|-------------|
| Need for PsySupp | 128 | 40.4 |
| Divided in: | | |
| “We have received psychological counselling/psychotherapy” | 29 | (22.7) |
| “We have received psychological counselling/psychotherapy partly” | 32 | (25.0) |
| “We have not received but we needed psychological counselling/psychotherapy” | 67 | (52.3) |
| No need for PsySupp | 189 | 59.6 |
| “We have not received and we do not need psychological counselling/psychotherapy” | | |

Table 2: Need for PsySupp in parents

| | Need for PsySupp | | No need for PsySupp | | n |
|---------------------------------------------------|------------------|-------------|---------------------|------|-----|
| | n | % | n | % | |
| Total | 128 | 40.4 | 189 | 59.6 | 317 |
| Diagnostic groups** | | | | | |
| dsd-XX-p-f | 43 | 33.6 | 85 | 66.4 | 128 |
| dsd-XY-p-f | 37 | 58.7 | 26 | 41.3 | 63 |
| dsd-XY-p-m | 40 | 38.5 | 64 | 61.5 | 104 |
| dsd-XY-c-f | 8 | 38.1 | 13 | 61.9 | 21 |
| Main diagnoses** | | | | | |
| pAIS | 13 | 65.0 | 7 | 35.0 | 20 |
| cAIS | 5 | 41.7 | 7 | 58.3 | 12 |
| Partial/mixed gonadal dysgenesis | 26 | 54.2 | 22 | 45.8 | 48 |
| complete gonadal dysgenesis | 3 | 37.5 | 5 | 62.5 | 8 |
| Disorders of androgen synthesis | 11 | 50.0 | 11 | 50.0 | 22 |
| CAH | 42 | 33.9 | 82 | 66.1 | 124 |
| Severe hypospadias | 13 | 26.5 | 36 | 73.5 | 49 |
| other | 15 | 44.1 | 19 | 55.9 | 24 |
| Surgeries | | | | | |
| Gonadectomy* | 36 | 52.2 | 33 | 47.8 | 69 |
| Investigations in context of the diagnosis | | | | | |
| Taking a picture** | 70 | 49.0 | 73 | 51.0 | 143 |
| Radiography* | 63 | 47.7 | 69 | 52.3 | 132 |
| Laparoscopy** | 16 | 66.7 | 8 | 33.3 | 24 |
| Gonadal biopsy*** | 38 | 56.7 | 29 | 43.3 | 67 |
| Induction of puberty** | | | | | |
| Hormonal induction - received | 15 | 50.0 | 15 | 50.0 | 30 |
| Hormonal induction - planned | 28 | 58.3 | 20 | 41.7 | 48 |
| No hormonal introduction | 63 | 35.0 | 117 | 65.0 | 180 |

only significant results are stated: *p<0.05; **p<0.01; ***p<0.001

Table 4: Significant results of the study

Conclusion:

- The special situation of having a child with dsd is associated with a high need for PsySupp in parents.
- In particular, parents of children with XY dsd with androgen effects other than hypospadias expressed a high need of PsySupp.
- There was no significant association between parents' perception of their child's genitalia and need for PsySupp.
- Gonadectomy, induction of puberty and different investigations in context of the diagnosis are associated with need for PsySupp in parents.
- PsySupp for parents should be an obligatory part of interdisciplinary care of children with dsd to reduce fears and concerns regarding gender issues, hormonal treatment and future surgery.