

How do Individuals with Complete Androgen Insensitivity Syndrome, Mayer-Rokitansky-Küster-Hauser Syndrome or Polycystic Ovary Syndrome Experience Contact to Other Affected Persons?

Wie erleben Personen mit kompletter Androgenresistenz, Mayer-Rokitansky-Küster-Hauser Syndrom und polyzystischem Ovarialsyndrom den Kontakt zu anderen Betroffenen?

Authors

K. Krupp¹, M. Fliegner¹, F. Brunner¹, S. Brucker², K. Rall², H. Richter-Appelt¹

Affiliations

¹ Institute for Sex Research and Forensic Psychiatry, University Medical Center Hamburg-Eppendorf, Hamburg
² Department of Obstetrics and Gynaecology, Tübingen University Hospital, Tübingen

Key words

- Müllerian agenesis
- PCOS (polycystic ovary syndrome)
- CAIS (complete androgen insensitivity syndrome)
- support group

Schlüsselwörter

- MRKH-Syndrom (Mayer-Rokitansky-Küster-Hauser)
- PCOS (polyzystisches Ovarialsyndrom)
- CAIS (komplette Androgenresistenz)
- Selbsthilfe

received 18. 7. 2012
 revised 18. 9. 2012
 accepted 17. 10. 2012

Bibliography

DOI <http://dx.doi.org/10.1055/s-0032-1327979>
 Geburtsh Frauenheilk 2012; 72: 1009–1017 © Georg Thieme Verlag KG Stuttgart · New York · ISSN 0016-5751

Correspondence

Prof. Dr. Hertha Richter-Appelt
 University Medical Center Hamburg-Eppendorf
 Institute for Sex Research and Forensic Psychiatry
 Martinistraße 52
 20246 Hamburg
 hrichter@uke.de

Abstract

Persons with different sex characteristics may suffer from a feeling of being “different” or “not normal”. In this study, persons with one of 3 diagnoses (complete androgen insensitivity syndrome [CAIS]; Mayer-Rokitansky-Küster-Hauser syndrome [MRKHS], polycystic ovary syndrome [PCOS]) were asked whether they had contact to other affected persons and how they assessed this contact. The correlation between contact and psychological distress was evaluated.

Material and Methods: Information on contacts to other affected individuals was obtained using a written questionnaire. Psychological distress was measured using the German version of the BSI (Brief Symptom Inventory).

Results: Data from 11 individuals with CAIS, 49 women with MRKHS and 55 women with PCOS was analysed. The frequency of contacts to other affected individuals differed between the different diagnostic groups (with the highest frequency reported for the group with CAIS, and the lowest for the group with PCOS). Overall, the majority of individuals considered such contacts beneficial (CAIS 81.8%; MRKHS 90%; PCOS 83.3%). The frequency of contacts and their assessment were not found to be correlated with psychological distress. The three diagnostic groups differed in the proportion of people who indicated a wish for contact with other affected persons. The desire to have contact with other affected persons was most commonly expressed by women with PCOS and high levels of psychological distress (60.9%).

Conclusion: Persons with different sex characteristics can benefit from contact to other affected individuals. Particularly women with PCOS and increased levels of psychological distress may benefit if the issue of support groups is addressed during treatment.

Zusammenfassung

Frauen, die von Veränderungen der Geschlechtsmerkmale betroffen sind, können unter dem Gefühl leiden, „anders“ oder „nicht normal“ zu sein. In der vorliegenden Arbeit wurden Personen aus 3 Diagnosegruppen (komplette Androgenresistenz, CAIS; Mayer-Rokitansky-Küster-Hauser Syndrom, MRKHS; polyzystisches Ovarialsyndrom, PCOS) befragt, ob sie Kontakt zu anderen Betroffenen hatten und wie sie diesen Kontakt bewerteten. Außerdem wurde untersucht, ob ein Zusammenhang zu psychischer Belastung bestand.

Material und Methoden: Im Rahmen eines schriftlichen Fragebogens wurden Angaben zum Kontakt mit anderen Betroffenen erhoben. Die psychische Belastung wurde über die deutsche Version des BSI (Brief Symptom Inventory) erfasst.

Ergebnisse: Die Antworten von 11 Personen mit CAIS, 49 Frauen mit MRKHS und 55 Frauen mit PCOS konnten ausgewertet werden. Die Häufigkeit des Kontakts zu anderen Betroffenen unterschied sich zwischen den Diagnosegruppen (am häufigsten in der Gruppe mit CAIS, am seltensten in der Gruppe mit PCOS). Insgesamt bewertete die Mehrheit diesen Kontakt als hilfreich (CAIS 81,8%; MRKHS 90%; PCOS 83,3%). Die Kontakthäufigkeit und die Bewertung hingen nicht mit der psychischen Belastung zusammen. Die 3 Diagnosegruppen unterschieden sich in dem Anteil der Personen, die sich Kontakt zu anderen wünschten. Am häufigsten äußerten diesen Wunsch Frauen mit PCOS, bei denen eine auffällige psychische Belastung vorlag (60,9%).

Schlussfolgerungen: Frauen, bei denen Veränderungen der Geschlechtsmerkmale vorliegen, können vom Kontakt zu anderen Betroffenen profitieren. Insbesondere für die Frauen mit PCOS, welche eine erhöhte psychische Belastung berichten, kann es hilfreich sein, in der Behandlung das Thema Selbsthilfe anzusprechen.

Introduction

Persons with different sexual characteristics often report a feeling of being “different from others” or of “not being normal”. This has been described both for individuals with different primary sexual characteristics and for those with different secondary sexual characteristics [1–3]. In such cases, contact to other affected individuals or attending support groups can be experienced as supportive and de-stigmatising [4]. However, the authors of this study did not find any published reports on how individuals with complete androgen insensitivity syndrome (CAIS), Mayer-Rokitansky-Küster-Hauser syndrome (MRKHS) or polycystic ovary syndrome (PCOS) experience contact to other affected individuals, what they find to be helpful, and whether some individuals felt that such contact also had negative aspects.

Description of the syndromes investigated

The investigated diagnostic groups consisted of syndromes which result in changes to female sexual characteristics or female sexual development and which are accompanied by infertility (CAIS and MRKHS) or reduced fertility (PCOS).

CAIS and MRKHS are rare disorders and are classified as belonging to the group of disorders of sex development, DSD [5,6].

The prevalence of CAIS is reported to be approximately 1 in 20000 live births with an 46,XY karyotype [7]. Despite the 46,XY karyotype, persons with CAIS are phenotypically female with external female genitalia due to complete insensitivity of cells to androgens. As the testes develop and release anti-Müllerian hormone, the uterus does not develop and the vagina ends blindly in a pouch. The diagnosis is frequently made in puberty because of primary amenorrhoea; breasts develop normally but pubic and underarm hair is either lacking or reduced [8]. The sexual identity of persons with CAIS is usually described as unambiguously female [8,9], however there are also other reports in the literature indicating an experienced sexual identity that differs from that of other women [10, 11]. Personal contacts with affected persons (when preparing the study) also showed that some did not feel the term “woman” to be a suitable description, which is why we have used gender-neutral terms to describe this group in our study.

MRKHS or Müllerian agenesis occurs in around 1 of 5000 female births [12]. It is characterised by agenesis of the uterus and vagina in women with an XX karyotype and is presumably of polygenetic origin. The ovaries are fully developed and functional [13]. Due to the undeveloped or extremely shortened vagina, vaginal sexual intercourse without previous medical treatment is not possible in most cases.

PCOS is one of the most common endocrine disorders in women of reproductive age; reports on its prevalence range from 5 to 17.8% (using the Rotterdam criteria), depending on the diagnostic criteria [14, 15]. In addition to hyperandrogenism and oligorrhoea or amenorrhoea, other external characteristics include acne, obesity, hirsutism and alopecia [16]. Some women with PCOS show a “masculinisation” of their external appearance.

What all of these diagnostic groups have in common are changes to certain female sexual characteristics or to their sexual development. However, the syndromes differ considerably with respect to the impact they have on the individual affected, particularly with regard to the wish to have children (persons with CAIS cannot have their own genetic children; in some countries, women with MRKHS may have genetic children if they enlist the help of a surrogate mother; women with PCOS are usually subfertile

but pregnancy is possible for some women with PCOS). The widely differing prevalence of the syndromes (CAIS and MRKHS are rare disorders, PCOS is relatively widespread) also needs to be taken into account when considering the results of this study.

Contact to other affected individuals

Contact to other affected persons can take very different forms. In addition to individual personal contacts, e.g. during a stay in hospital, individuals may also be in contact with a group (group therapy, various support groups; cf. also [17,18]). Contacts increasingly also come about via the internet, in the form of private e-mail contacts or discussion forums (cf. [19,20]).

The central task of self-help groups has been described as the provision of social and emotional support as well as the provision of information [21].

Overall, contact to self-help/support groups has been experienced differently; some people find it helpful, others reject it [2, 22,37].

For people with CAIS and PCOS it has been reported that the experience of “otherness” or isolation associated with the diagnosis can be reduced through contact to other affected persons [2,21]. For intersex persons in particular, the importance of meeting “similar others” for the development of a positive identity was emphasised [23]. Our search of the literature did not find any comparable studies on self-help groups or contacts to other affected persons for persons with MRKHS. However, results obtained from an interdisciplinary Patient Day indicated that women with MRKHS also judged the exchange of experiences with other affected persons to be positive [20].

As providing information above and beyond the respective individual diagnosis is also an important task of support groups, this study also looks at how participants evaluated the information on various aspects of the disorder that was provided by their treating physician and whether this was related to their contact to other affected persons.

Psychological distress

There are a number of studies on psychological distress in persons with CAIS, MRKHS and PCOS, some of which point to increased levels of distress. However, the results are somewhat contradictory [24]. Studies have reported increased psychological distress but also completely unremarkable levels of distress in individual groups (CAIS: [25,26], MRKHS: [27–29], PCOS [30, 31]).

Individual results appear to indicate that persons in support groups cannot be taken as representative for the patient group as a whole [32]. One of the hypotheses for this is that persons with particularly high levels of distress are more likely to seek contact to other affected individuals or support groups. To assess this more precisely, our study recorded the levels of psychological distress of the persons taking part in the study. The study then examined whether there was any correlation between the level of psychological distress and contact to other affected persons (as well as how this contact was rated).

Aim of this study

This study aimed to examine contact to other affected persons with CAIS, MRKHS and PCOS and its possible correlation with psychological distress. In addition to the frequency of such contacts for the individual diagnostic groups, the study also aimed to investigate whether such contacts were considered helpful and whether contact to others was desired. Accordingly, the fol-

lowing null hypotheses were proposed for the study: the 3 diagnostic groups do not differ from each other with regard to levels of psychological distress, frequency of contact, the assessment of this contact and the desire for contact. The frequency of contact to other affected persons, the assessment of this contact and the desire for contact are not related to the level of psychological distress; the information received from the treating physician also has no impact on these variables.

Method



Data collection and participants

Data was collected between March 2010 and July 2011 as part of a research project on “Androgens quality of life and femininity in persons with CAIS, MRKHS and PCOS” at the University Clinic Hamburg-Eppendorf (supported by the Else Kröner Fresenius Stiftung; project leader: Prof. Dr. Richter-Appelt; the project was approved by the Ethics Commission of the local medical association). Participants (minimum age: 18 years) were recruited from all over Germany through their physicians and the hospitals where they received treatment (primarily through the Gynaecological University Hospital of Tübingen), through support groups (primarily the support group “Intersexuelle Menschen e.V.”), through the project’s website and with the help of appeals launched in professional journals. Participation consisted of filling out a questionnaire which had been compiled from standardised instruments and answering questions which the investigators developed themselves however space was also provided for the participants’ own comments. Confirmation of the respective diagnoses was conducted in 2 stages. The data given in the questionnaires was checked for plausibility and for its agreement with the diagnosis participants stated they had received. In addition, the participant’s physician was contacted and asked to confirm the diagnosis as well as to provide additional medical findings (the precondition for this was that participants authorised their physician to release their medical records). Questionnaires which could not be clearly classified into one of the diagnostic groups were excluded from the analysis. Questionnaires which had been filled out but did not include the participant’s consent to take part in the study were also excluded.

For a better comparison of certain variables (relationship status, occupation), additional data from a non-clinical sample (n = 932 women) was also reviewed; this data had been collected in the form of an online survey conducted as part of the research project.

Questionnaire

Information about contact to other affected persons was collected using questions developed by the researchers themselves. Recorded data included information as to whether such contacts had taken place, whether the contact was considered helpful and – if there had been no contact – whether the participant desired such contact. The last two questions included space for comments by the participants themselves.

Participants were also asked whether they felt that they had been given sufficient information by their treating physician about the diagnostics used, treatment, the consequences of the diagnosis and long-term consequences. An index (0–4) was compiled from the answers to these 4 points which showed how well the study participants felt they had been informed overall (0 = not sufficiently on any point; 4 = well informed on all points).

Psychological distress was assessed using the Brief Symptom Inventory (BSI, German version [33]). The questionnaire recorded individual psychological distress over the past week using 53 different items. The participant rated the extent to which they felt impaired by various medical conditions on a 5-point Likert scale (0 = not at all; 4 = very strongly). The answers were summarised in 9 scales (Somatisation, Obsessive-Compulsive, Interpersonal Sensitivity, Depression, Anxiety, Hostility, Phobic Anxiety, Paranoid Ideation, Psychoticism). In addition, an overall score was calculated which reflected the overall level of psychological distress.

The resulting values were converted into standardised T-values using the standard values given in the manual [33], which permitted a direct comparison to be made with the non-clinical reference samples (mean T-value for distribution is 50, standard deviation is 10. A value of 50 in a patient sample would thus correspond exactly to the mean value of the non-clinical reference group; a value of 70 would be 2 standard deviations above the mean value of the reference group and would thus point to markedly higher levels of distress).

In accordance with the BSI manual, persons who had T-values of 63 or more in their overall score or in at least 2 subscales were classified as “clinical cases” with conspicuously higher levels of psychological distress.

Statistical analysis

Statistical analysis was conducted using the PASW 18 software (SPSS). χ^2 -tests for 2×2 and 3×2 cross-classified tables were used to calculate the relationship between categorical variables. If statistical requirements were not met (expected frequency of individual cells < 5), exact χ^2 -tests (Fisher-Yates test) were used for calculation. Differences between groups were compared using Mann-Whitney U-test.

The results of the Brief Symptom Inventory (BSI) were recorded as standardised T-values, which were calculated based on the reference data of the test manual [35] (mean distribution of the T-values is 50, standard deviation is 10).

Any additional comments by participants were assessed qualitatively. Frequencies of the issues mentioned were calculated.

Results



Description of samples

A total of 126 questionnaires were completed, of which 5 had to be excluded due to formal faults (no consent given for participation in the study). Six more questionnaires were excluded due to missing or inappropriate diagnosis (no clear diagnosis of CAIS, MRKHS or PCOS). Thus, the data of 115 persons was included in the final evaluation (11 persons with CAIS; 49 women with MRKHS; 55 women with PCOS).

The study participants heard about the research project from a number of different sources. In the group with CAIS, the majority (63.6%; 7/11) heard about it through their support groups. In the MRKHS group, this proportion was distinctly lower (2%; 1/49), most persons in this group (65.3%; 32 of 49) were advised of the study by the hospital they attended or by their general practitioner. In the PCOS group, 78.2% (43/55) heard about the study from the hospital they attended or from the physician treating them on an outpatient basis; only one person (1.8%) was informed about the study by her support group. A detailed summary is given in [Table 1](#).

Table 1 How did study participants hear about the research project?

How did you hear of our study?	CAIS (n = 11)		MRKHS (n = 49)		PCOS (n = 55)	
	N	%	N	%	n	%
University clinic/specialised clinic/centre	1	9.1%	28	57.1%*	31	56.4%
Gynaecologist	–	–	2	4.1%	5	9.1%
Another physician	–	–	2	4.1%	7	12.7%
Support group	7	63.6%	1	2.0%	1	1.8%
Another affected person	1	9.1%	2	4.1%	1	1.8%
Internet	1	9.1%	6	12.2%	2	3.6%
Other	1	9.1%	–	–	1	1.8%
Not specified	–	–	8	16.3%	7	12.7%

* 25 women with MRKHS (51%) heard of the study through their treatment at Tübingen University Hospital.

The average age of participants with CAIS was 38.7 years (± 9.6); the average age of participants with MRKHS was 23.6 years (± 5.8) and that of participants with PCOS was 29.1 years (± 4.2). At the time of the study, 54.5% of the participants with CAIS were living with a partner (5 with a male partner, 1 with a female partner). In the MRKHS group, 75.5% were in a relationship, and 87.3% of the PCOS group were in a relationship. The proportion of persons living with a partner differed significantly between the diagnostic groups (Fisher-Yates test: p-value = 0.037) (reference value for the non-clinical sample: 73% living with a partner).

All participants with CAIS were either working or in education or training. In the MRKHS group, 45 persons (91.8%) were either working or in education or training (or at school/university), 4 (8.2%) were unemployed. Of the participants with PCOS, 40 (72.7%) were working or in education or training, 7 women (12.7%) were on maternity leave or had taken parental leave, 3 (5.5%) described themselves as housewives, 4 persons (7.3%) were unemployed, and 1 (1.8%) was incapable of working (reference value for the non-clinical sample: 71.5% employed or in education or training; 3.3% unemployed; 2.5% incapable of working or had taken early retirement for invalidity; 22.6% other).

Psychological distress

In accordance with the definition of the BSI for “clinical levels of psychological distress”, 6 persons (54.5%) in the study group with CAIS were classified as suffering from higher levels of psychological distress. In the MRKHS group the BSI could only be evaluated for 48 participants; of these 48 participants, 26 (54.2%) were considered to have higher levels of psychological distress. In the PCOS group, 29 persons (52.7%) showed higher levels of psychological distress. The distribution between the diagnostic groups did not differ significantly ($\chi^2 = 0.027$; df = 2; p-value = 0.987).

The results for the overall BSI value (GSI, Global Severity Index) and the 9 subscales for each diagnostic group are given in **Table 2**.

Contact to other affected persons

All of the study participants with CAIS reported that they were in contact with other affected persons. In the MRKHS group, only 48 women answered this question; of these, 30 (62.5%) had contact to other affected persons. Among the group with PCOS only 6 persons (10.9%) were in contact with other affected women. The proportion of people with contact to other affected persons depended significantly on the respective diagnosis (Fisher-Yates test: p-value < 0.001).

Table 2 BSI results (T-values^a) for each diagnostic group.

	CAIS (n = 11)		MRKHS (n = 48 ^b)		PCOS (n = 55)	
	MW	(SD)	MW	(SD)	MW	(SD)
Overall GSI score	60.91	(10.30)	57.25	(14.15)	59.04	(12.98)
Subscales						
Somatisation	56.73	(11.16)	52.90	(10.66)	54.35	(11.11)
Obsessive-Compulsive	59.36	(9.78)	54.21	(11.82)	55.84	(10.76)
Interpersonal Sensitivity	61.64	(10.46)	58.06	(11.48)	59.65	(11.09)
Depression	60.36	(12.38)	55.56	(13.18)	58.98	(12.49)
Anxiety	57.27	(11.22)	53.33	(12.31)	53.95	(11.76)
Hostility	60.09	(11.20)	59.50	(10.83)	59.87	(9.85)
Phobic Anxiety	56.64	(12.46)	54.31	(11.42)	53.02	(10.90)
Paranoid Ideation	54.00	(11.14)	57.29	(11.52)	56.85	(15.56)
Psychoticism	59.91	(11.31)	56.63	(12.02)	56.16	(10.89)
Clinical cases^c	n	(%)	n	(%)	n	(%)
	6	54.5%	26	54.2%	29	52.7%

^a Mean = 50, standard deviation = 10

^b The BSI results could not be calculated for one of the persons with MRKHS due to a lack of data.

^c Participants (number and percentage), who fulfilled the criteria for “clinically relevant level of psychological distress”: GSI T-value ≥ 63 , or T-value ≥ 63 in 2 or more subscales).

Table 3 Reasons given for the respective answer: contact helpful yes/no.

Contact to other affected persons ...	
Category	Typical comment (original text)
Was helpful because:	
Feeling of not being alone	"You find out that you are not alone with the diagnosis"
Similar experience/more understanding	"You have someone who has gone through the same things as you have yourself. That is incredibly helpful."
Exchange of experience/information	"Was able to find out about surgery from them"
Psychological support	"I felt/feel it to be a place where someone is there for me on bad days."
Changed the way of dealing with the diagnosis	"Thanks to the forum I can now talk more openly about it"
Able to show weakness	"You realise that you can sometimes allow yourself to show weakness"
Was not helpful because:	
The focus was on negative aspects	"However, I feel one should not only occupy oneself with the diagnosis and the problems. At one stage this 'continual rehashing of problems' with other affected persons got to be too much for me."
Contact is stressful	"At the moment it is more stressful, because I experience it as very inflexible and it does not help me progress further"
Had other expectations	"I was too unrealistic"
Practical reasons	"Unfortunately because of the distance, benefit only limited"

Table 4 Reasons given for the respective answer: contact desired yes/no.

Contact to other affected persons ...	
Category	Typical comment
Is desired because:	
Hear about other experiences/exchange	"Would like to hear about others' experience with it and how they deal with it"
More information	"Want to find out more about this disease"
Is not desired because:	
Not felt to be necessary	"I can deal with it quite well"
Contact experienced as stressful	"Contact would be too stressful, real."
Do not want to think about diagnosis	Partly try not to think about it"

Was contact to others rated as helpful?

When study participants did have contact to other affected persons, the majority (87.2%) of the participants experienced such contact as helpful. Of the participants with CAIS who had contact to other affected persons, 81.8% (9/11) experienced this contact as helpful. In the group with MKRHS the percentage of women who felt contact was helpful was 90% (27/30); in the group with PCOS this figure was 83.3% (5/6). The percentage did not correlate significantly with the individual diagnosis (Fisher-Yates test: p -value = 0.596).

A total of 41 participants in the study included additional comments on how they assessed contact with other affected persons. People who rated contact as helpful most frequently cited the experience of not being alone (13 mentions) as their reason for such contact. The exchange of experience and information on the diagnosis were also listed as important (6 mentions), followed by psychological support and the greater understanding they received from other affected persons who had had similar experiences (5 mentions each). Other comments on the benefits of contact included being able to talk openly about the diagnosis (1 mention) and being able to let one's guard down in a group of other persons with the diagnosis (1 mention).

If the contact was not rated as beneficial, the reasons for this, cited in 3 cases, included the focus on negative topics in the support group, while 2 cases experienced contacts to other affected persons as stressful. One person with CAIS reported a feeling of "not belonging" and of exclusion even when meeting other persons with a diagnosis of CAIS. Other expectations and practical reasons (distance from the place of residence) were also cited as explanations for why the contact was not felt to be helpful.

Typical comments for the different categories are listed in **Table 3**.

Do people wish for contact with other affected persons?

Individuals who had no contact with other affected persons were asked whether they wished for such contact. In the group with MRKHS 52.9% (9 of 17) wished for such contact, in the PCOS group 39.6% (19 of 48) wished for such contact. This distribution did not differ significantly between the 3 diagnostic groups (Fisher-Yates test: p -value = 0.561).

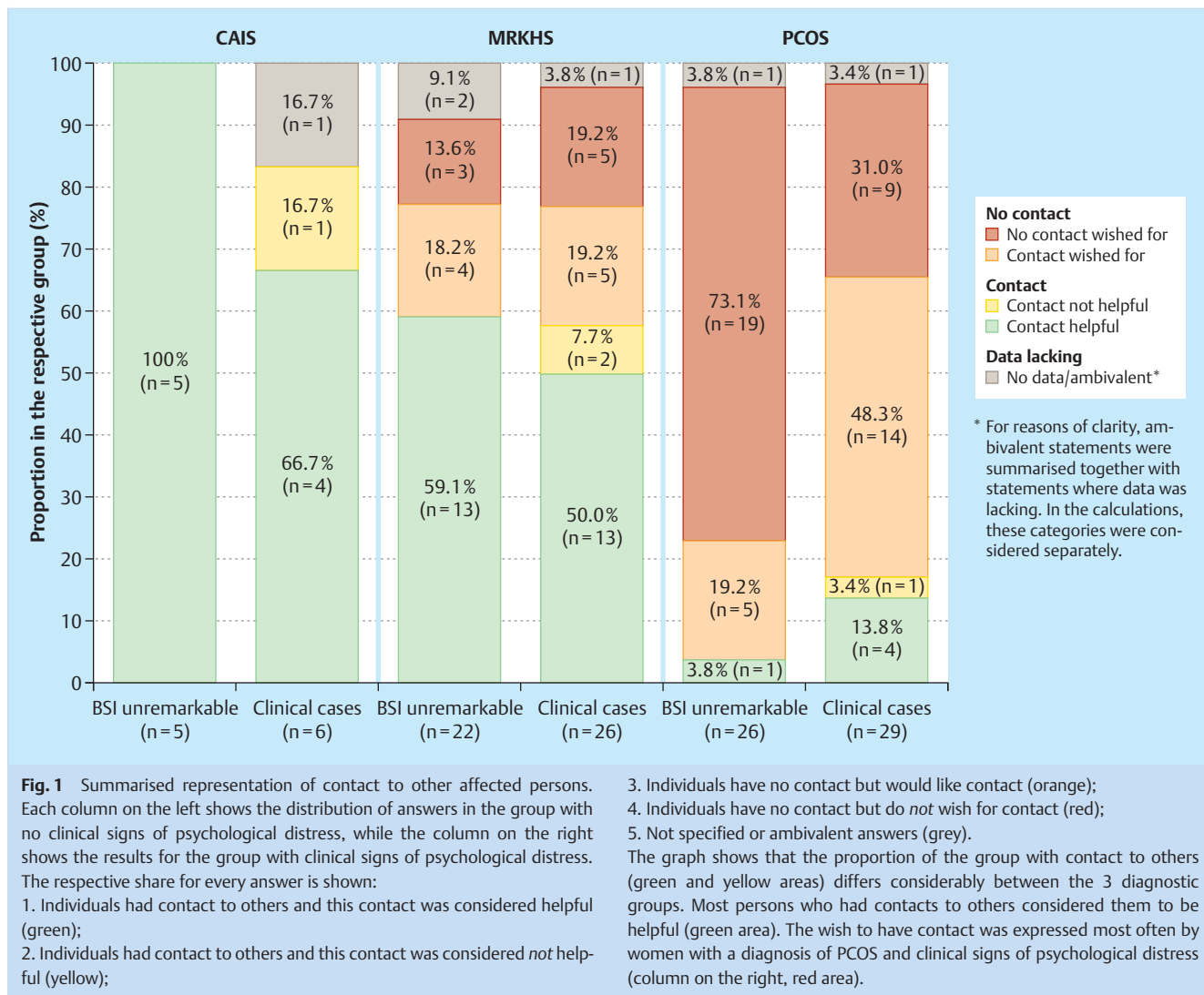
Thirty persons appended additional comments regarding their wish for contact to other affected persons. If contact was desired, 9 persons stated their reason as wanting to hear about others' experience of the diagnosis and wishing to exchange experiences. One woman with PCOS hoped to obtain more information about her diagnosis.

Persons who did not wish for contact to other affected persons usually stated that they did not feel it to be necessary for them (6 mentions). Three women rejected such contact because they did not want to be reminded of their diagnosis. Two women with MRKHS did not want any contact because they considered this would be too distressing and were afraid that discussions would focus too much on negative topics.

Table 4 gives a summary of typical comments on the respective topics.

Correlation between contacts to other affected persons and psychological distress

In the overall group, no significant correlation was found between higher levels of psychological distress (based on the crite-



ria for “clinical cases”) and contact to other affected persons (analysis of the 3 diagnostic groups taken together; $\chi^2=0.365$; $df=1$; $p\text{-value}=0.546$). This also applied when the diagnostic groups were analysed separately (CAIS: no calculation, all individuals have contact to others; MRKHS: $\chi^2=0.065$; $df=1$; $p\text{-value}=0.798$; PCOS: Fisher-Yates test: $p\text{-value}=0.197$).

Similarly, in the group which had contact to other affected persons, no correlation was found between the level of psychological distress and evaluation of the contact as helpful (analysis of all 3 diagnostic groups taken together; $\chi^2=3.375$; $df=2$; $p\text{-value}=0.185$).

This also applied to the individual diagnostic groups (CAIS: Fisher-Yates test: $p\text{-value}=1.000$; MRKHS: Fisher-Yates test: $p\text{-value}=0.483$; PCOS: Fisher-Yates test: $p\text{-value}=1.000$).

For the overall group, the wish to have contact with other affected persons depended significantly on the level of psychological distress (Fisher-Yates test: $p\text{-value}=0.024$). In the group of clinically distressed persons 57.6% (19/33) wished for contact, while the wish for contact with other affected persons was distinctly lower in the group with clinically unremarkable levels of distress (28.1%; 9/32). The following correlations were found for the different diagnostic groups. This calculation was not made for the CAIS group as all persons in the group had contacts to other

affected persons. There was no correlation in the group with MRKHS (Fisher-Yates test: $p\text{-value}=1.000$). In the group with PCOS the level of psychological distress was found to correlate significantly with the wish for contact to other affected persons (Fisher-Yates test: $p\text{-value}=0.007$). Of the women with clinically significant levels of psychological stress (“clinical cases”), 60.9% (14/23) wished to have contact to other affected persons, while in the group without clinically relevant levels of distress, only 20% (5/25) expressed this wish.

A summary of the results is given in **Fig. 1**.

Information obtained from the treating physician

How well individuals felt that their physician had informed them differed significantly between the diagnostic groups (Fisher-Yates test: $p\text{-value}<0.001$). Overall, the extent of information provided by the physician was rated highest by the MRKHS group (median = 4), and lowest by the CAIS group (median = 0). With a median value of 3, the rating given by the PCOS group was between that of the 2 other groups.

No difference was found between persons who had contact to other affected persons and those who did not with regard to their statements about the extent of the information provided by their physician ($U=1442$; $p\text{-value}=0.426$). There was also no differ-

ence with regard to the information provided by their physician between persons who considered contact to be helpful and those who did not ($U = 43.5$; $p\text{-value} = 0.112$; however it should be noted here that the group of persons who did not consider contact to be helpful was very small, with only 4 persons). Similarly, no difference in the assessment of the extent to which information was provided by the physician was found between the group who wished for contact compared to the group of persons who did not ($U = 478.5$; $p\text{-value} = 0.718$).

The level of psychological distress (BSI overall value) did also not correlate with the patients' assessment of the extent of information provided by their physicians.

Discussion

Central results and comparison with other studies

The null hypotheses formulated at the beginning regarding the level of psychological distress, the assessment of contact to affected persons as being helpful, and the wish to have contacts to other affected persons were not rejected by the results of the study. The 3 diagnostic groups did not differ with regard to these variables. The null hypothesis regarding the frequency of contact must be rejected as the 3 groups differed significantly from each other. In the sample investigated, all persons with CAIS had contacts to other affected persons with CAIS, in the group of women with MRKHS the majority reported being in contact with other persons with MRKHS while, in comparison, in the group of women with PCOS a much lower proportion of women reported being in contact with other women with PCOS. Of the 3 diagnostic groups investigated, PCOS is the most common; overall more women are diagnosed with this syndrome. Given the fact that the incidence of women with PCOS is higher, one could conjecture that women with PCOS would also have more opportunity to establish contact to other affected women or to join support groups, as the choices available for this fairly common diagnosis are greater. The existing data however tell a rather different story. When interpreting the results it must be borne in mind that the participants recruited heard about the study in many different ways. Persons with CAIS were primarily informed about the study through the offices of support groups for XY women; accordingly, they were the group which most frequently cited having contact to other affected persons. We can safely assume that the results for the group with CAIS are not representative and that the percentage of persons with contact to other persons with the same diagnosis is probably overestimated based on the sample of people in our study. To compare these results, we looked at the Hamburg study on intersexuality; in this study 50% of participants reported that they had some experience of support groups [34]. For women with PCOS another study cited a figure of 51.5% (17/33) of persons who had described attending a support group [31]. Since in our study, participants were partly recruited through the nationwide network of PCOS support groups, it can be assumed that the percentage of women with PCOS who attend support groups was overestimated. The percentage of persons with a particular diagnosis who have contact to other affected persons or to support groups would have to be investigated in larger, more representative studies. The ways in which potential study participants are recruited play a very important role.

A correlation between the level of psychological distress and the desire to have contact with other affected persons was only

found in the group with PCOS; the desire to have contact was voiced more frequently by women with higher levels of psychological distress. There were no differences between the diagnostic groups with regard to the frequency of such contact or the assessment of contact.

In the sample investigated, there was no indication that primarily persons with particularly high levels of psychological distress sought contact to other persons with the same diagnosis or to support groups. This is in accordance with the results of Jauca et al. [31] who, in their study of 33 women with PCOS, also did not report any significant differences with regard to the level of psychological distress between persons who attended support groups and those who did not.

The diagnostic groups did differ in their assessment of the information provided by their attending physician. The group of women with MRKHS reported the highest levels of satisfaction with the extent of information they had received, followed by the group of women with PCOS; persons with CAIS reported the lowest level. No correlation was found between the extent of information received from their attending physician and the frequency of contact, the assessment of such contact, or the desire for contact.

Using the examples 'androgen insensitivity' and 'adrenogenital syndrome' (AGS, one of a group of intersex conditions) Warne reported that support groups went through various phases during which the focus of the group changed (from obtaining information and supporting other members of the group to lobbying for social change) [35]. In our study, contact to other affected persons were investigated in a more general sense (not only contact to support groups), and it was therefore not possible to make any statements about changing phases within groups. It is conceivable however that the backdrop to the assessment of contacts as "not helpful" by individual persons could be a lack of fit between the needs of the individual and the current phase of the group.

In this context it is important to point out again the differences between the 3 diagnostic groups investigated, particularly with respect to the potential to have genetic children of one's own. Women with PCOS can become pregnant, whereas persons with CAIS and MRKHS cannot. Depending on the phase of life they are in, becoming pregnant and the medical support they may require could play a key role for women with PCOS. This would probably also be reflected in the topics participants put forward for discussion in support groups. Which topics individual persons in support groups wish to talk about and which areas they wish to learn more about were not investigated in this study. Future studies could investigate which topics persons in support groups wish (or do not wish) to talk about and which topics they experience as particularly helpful, depending on their current circumstances, for the different diagnostic groups.

In a report which included her own experiences, an affected physician described her contacts to an AGS action group of affected persons and emphasised the importance of the different generations within the support group [36]. Medical advances and changes in the problems experienced by patients mean that it is necessary to look at self-help groups over 10 to 20-year periods [36]. This aspect was not included in our study and could be investigated in more detail in subsequent studies: Did the participant have any contact to persons of similar age or going through the same stage of treatment? Was contact to "experienced" persons with the same diagnosis felt to be particularly helpful? Or could the experiences of older affected persons not be compared

to the individual's own situation because the treatment available had changed?

The reasons given for the assessment of contact to others listed above more or less correspond to the range of topics described in a qualitative study on support groups for PCOS [21] with the difference that, in our study, negative aspects were also mentioned (e.g. the stress experienced through contact to other affected persons).

Overall, it was noticeable that the percentage of persons with higher levels of psychological distress in all 3 diagnostic groups was more than 50% and thus relatively high. These figures correspond to the results of the Hamburg Intersex Study [25]; in the Hamburg study, 59% of participants fulfilled the criteria for clinically relevant levels of distress. Jauca et al. described similar results for women with PCOS [31]; they reported a figure of 42.4% for persons with clinically higher levels of psychological distress. This once again highlights the importance of ancillary diagnostic investigations in persons with different sexual characteristics as these could help identify patients with high levels of psychological distress and provide them with support or therapy.

Limitations

As already discussed above, the manner in which participants were contacted and recruited into the study plays an important role for the interpretation of our results. Most of the participants with CAIS heard about the study through a support group; we therefore have to assume that their results regarding the frequency of contact with other affected persons are skewed. Due to the way in which we chose to recruit participants into the study (not a random sample, only persons interested in taking part in the study were included), we cannot assume that the results are representative for any of the 3 groups. The results reflect the perceptions of some persons with these diagnoses and should not be generalised. They offer a first insight into the subject matter and provide starting points for further investigations. The differentiated examination of the proband sample resulted in a number of subgroups, some of which were quite small. Thus, despite the initially large sample group, this may have resulted in correlations between individual variables not being visible in the results.

This study did not investigate the differences between the 3 syndromes in detail; in particular, the differing importance for affected persons was not examined. Only one specific aspect was highlighted here. The data described here gives a first overview of contact to other affected persons with CAIS, MRKHS and PCOS. To assess individual impact factors in more detail and potentially make recommendations for different patient groups, the following points need to be investigated in further studies: In which context did contact take place? For how long? With regard to support groups: How was the group organised? What goals did the group have? What were the expectations on joining a support group?

Conclusion

The majority of participants in our study reported that contact to other affected persons was helpful; however, no correlation between contacts and the level of psychological distress was found in any of the diagnostic groups. The current data do not permit any recommendations to be made on which persons would benefit from contact to a support group and which would not. To formulate any such recommendation a further study would be necessary to differentiate between the types of contact and examine

them in connection with the specific characteristics of affected persons.

It should be noted, however, that among the women with PCOS, those with clinically relevant levels of psychological distress more frequently expressed the wish to have contact with other affected persons. This group could benefit from information on what services are available. However, it should be noted that some participants also reported negative experiences associated with contacts to other affected persons. This issue also needs to be broached when counselling patients.

Practical implications

Some women with different sexual characteristics report that they benefit from contact to other affected individuals. In particular women with PCOS and clinically relevant levels of psychological distress expressed more frequently the wish to have contact to others. For this group, it could be beneficial to address the topic of self-help groups during treatment and to advise patients of suitable groups available to them.

Acknowledgement

We would like to thank everyone who was involved in the study; our particular thanks go to the people who participated in the study who were prepared to share their experiences with us. We would like to thank the Else Kröner Fresenius Stiftung for financially supporting this research project.

Conflict of Interest

None.

References

- 1 Heller-Boersma JG, Edmonds DK, Schmidt UH. A cognitive behavioural model and therapy for utero-vaginal agenesis (Mayer-Rokitansky-Küster-Hauser syndrome: MRKH). *Behav Cogn Psychoth* 2009; 37: 449–467
- 2 Garrett CC, Kirkman M. Being an XY female: an analysis of accounts from the website of the androgen insensitivity syndrome support group. *Health Care Women Int* 2009; 30: 428–446
- 3 Kitzinger C, Willmott J. "The thief of womanhood": women's experience of polycystic ovarian syndrome. *Soc Sci Med* 2002; 54: 349–361
- 4 Richter-Appelt H. Psychologische und psychotherapeutische Interventionen. In: Schweizer K, Richter-Appelt H, Hrsg. *Intersexualität kontrovers: Grundlagen, Erfahrungen, Positionen*. Gießen: Psychosozial-Verlag; 2012: 355–369
- 5 Schweizer K. Sprache und Begrifflichkeiten. *Intersexualität benennen*. In: Schweizer K, Richter-Appelt H, Hrsg. *Intersexualität kontrovers: Grundlagen, Erfahrungen, Positionen*. Gießen: Psychosozial-Verlag; 2012: 19–39
- 6 Hughes IA, Houk C, Ahmed SF et al. Consensus statement on management of intersex disorders. *Arch Dis Child* 2006; 91: 554–563
- 7 Blackless M, Charuvastra A, Derrtyck A et al. How sexually dimorphic are we? Review and synthesis. *Am J Hum Biol* 2000; 12: 151–166
- 8 Richter-Appelt H. *Intersexualität – Störungen der Geschlechtsentwicklung*. Bundesgesundheitsbl – Gesundheitsforsch – Gesundheitsschutz 2007; 50: 52–61
- 9 Oakes MB, Eyvazzadeh AD, Quint E et al. Complete androgen insensitivity syndrome – a review. *J Pediatr Adolesc Gynecol* 2008; 21: 305–310
- 10 Brunner F, Prochnow C, Schweizer K et al. Körper- und Geschlechtererleben bei Personen mit kompletter Androgeninsensitivität. *Z Sexualforsch* 2012; 25: 26–48

- 11 Handford C, Brunner F, Schweizer K et al. Brauchen wir ein drittes Geschlecht? Erwachsene mit Androgeninsensitivität nehmen Stellung. In: Schweizer K, Richter-Appelt H, Hrsg. Intersexualität kontrovers: Grundlagen, Erfahrungen, Positionen. Gießen: Psychosozial-Verlag; 2012: 429–445
- 12 Aittomaki K, Eroila H, Kajanoja P. A population-based study of the incidence of Mullerian aplasia in Finland. *Fertil Steril* 2001; 76: 624–625
- 13 American College of Obstetricians and Gynecologists. ACOG Committee Opinion No. 355: Vaginal agenesis: diagnosis, management, and routine care. *Obstet Gynecol* 2006; 108: 1605–1609
- 14 Azziz R, Woods KS, Reyna R et al. The prevalence and features of the polycystic ovary syndrome in an unselected population. *J Clin Endocrinol Metab* 2004; 89: 2745–2749
- 15 March WA, Moore VM, Willson KJ et al. The prevalence of polycystic ovary syndrome in a community sample assessed under contrasting diagnostic criteria. *Hum Reprod* 2010; 25: 544–551
- 16 Rosenfield RL. What every physician should know about polycystic ovary syndrome. *Dermatol Ther* 2008; 21: 354–361
- 17 Cohen S, Underwood L, Gottlieb BH et al. Social Support Measurement and Intervention: a Guide for Health and social Scientists. Oxford, New York: Oxford University Press; 2000
- 18 Trojan A, Kofahl C. Selbsthilfe, Selbsthilfegruppen und Selbsthilfeförderung. In: BZgA, Hrsg. Gamburg: Verlag für Gesundheitsförderung; 2011: 491–496
- 19 Eysenbach G, Powell J, Englesakis M et al. Health related virtual communities and electronic support groups: systematic review of the effects of online peer to peer interactions. *Brit Med J* 2004; 328: 1166–1170
- 20 Rall K, Wallwiener M, Rießel S et al. Optimierung von Aufklärung, Betreuung und Compliance bei Patientinnen mit seltenem gynäkologischen Krankheitsbild – am Beispiel des Mayer-Rokitansky-Küster-Hauser-Syndroms (MRKH-Syndrom). *Geburtsh Frauenheilk* 2008; 68: 1102–1104
- 21 Percy CA, Gibbs T, Potter L et al. Nurse-led peer support group: experiences of women with polycystic ovary syndrome. *J Adv Nurs* 2009; 65: 2046–2055
- 22 Kirkman M. Thinking of something to say: public and private narratives of infertility. *Health Care Women In* 2001; 22: 523–535
- 23 Schweizer K. Identitätsbildung und Varianten der Geschlechtsidentität. In: Schweizer K, Richter-Appelt H, Hrsg. Intersexualität kontrovers: Grundlagen, Erfahrungen, Positionen. Gießen: Psychosozial-Verlag; 2012: 459–484
- 24 Krupp K, Fliegner M, Brunner F et al. Quality of life and psychological distress in women with Mayer-Rokitansky-Küster-Hauser syndrome and individuals with complete androgen insensitivity syndrome. *J Health Psychol* (submitted)
- 25 Schützmann K, Brinkmann L, Schacht M et al. Psychological distress, self-harming behavior, and suicidal tendencies in adults with disorders of sex development. *Arch Sex Behav* 2009; 38: 16–33
- 26 Hines M, Ahmed SF, Hughes IA. Psychological outcomes and gender-related development in complete androgen insensitivity syndrome. *Arch Sex Behav* 2003; 32: 93–101
- 27 Huber K, Hoffmann S, Richter-Appelt H. Das Mayer-Rokitansky-Küster-Hauser-Syndrom. Eine Übersicht über den Stand der internationalen psychosomatischen Forschung. *Z Sexualforsch* 2009; 22: 318–339
- 28 Laggari V, Diareme S, Christogiorgos S et al. Anxiety and depression in adolescents with polycystic ovary syndrome and Mayer-Rokitansky-Küster-Hauser syndrome. *J Psychosom Obst Gyn* 2009; 30: 83–88
- 29 Morgan EM, Quint EH. Assessment of sexual functioning, mental health, and life goals in women with vaginal agenesis. *Arch Sex Behav* 2006; 35: 607–618
- 30 Deeks AA, Gibson-Helm ME, Paul E et al. Is having polycystic ovary syndrome a predictor of poor psychological function including anxiety and depression? *Hum Reprod* 2011; 26: 1399–1407
- 31 Jauca R, Jäger S, Franke GH. Psychische Belastung, Lebenszufriedenheit und Krankheitsverarbeitung bei Frauen mit dem Polyzystischen Ovar-Syndrom (PCOS). *Z Med Psychol* 2010; 19: 38–47
- 32 Zucker KJ. Re: Sexual behaviors, sexual orientation and gender identity in adult intersexuals: a pilot study. *J Urology* 2002; 168: 1507–1508
- 33 Franke GH. Brief Symptom Inventory von L. R. Derogatis (Kurzform der SCL-90-R). Deutsche Version – BSI: Manual. Göttingen: Beltz Test; 2000
- 34 Schweizer K, Richter-Appelt H. Die Hamburger Studie zur Intersexualität. Ein Überblick. In: Schweizer K, Richter-Appelt H, Hrsg. Intersexualität kontrovers: Grundlagen, Erfahrungen, Positionen. Gießen: Psychosozial-Verlag; 2012: 187–205
- 35 Warne G. Support groups for CAH and AIS. *Endocrinologist* 2003; 13: 175–178
- 36 Kraus-Kinsky E. Adrenogenitales Syndrom. Persönliches Erleben zwischen eigener Lebensgeschichte und dem Dasein als Ärztin. In: Schweizer K, Richter-Appelt H, Hrsg. Intersexualität kontrovers: Grundlagen, Erfahrungen, Positionen. Gießen: Psychosozial-Verlag; 2012: 161–173
- 37 Erim Y, Beckmann M, Hoffmann O et al. Prädiktoren psychischer Belastungen bei Brustkrebspatientinnen – Worauf sollten Ärzte und Pflegekräfte in der onkologischen Praxis achten? *Geburtsh Frauenheilk* 2011; 71: 285–291

Deutschsprachige Zusatzinformationen online abrufbar unter:
www.thieme-connect.de/ejournals/toc/gebfra.