Psychological Distress in Women With Uterovaginal Agenesis (Mayer-Rokitansky-Küster-Hauser Syndrome, MRKH)

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Background: *Uterovaginal agenesis (Mayer-Rokitansky-Küster-Hauser Syndrome; MRKH) is a congenital nonformation of the vagina and the uterus, but with normal ovaries.* **Objective:** *The authors investigated the psychological impact of this disorder, about which very little is known.* **Method:** *A group of 66 women with MRKH were compared with 31 control-group women on a range of self-rating scales assessing psychological distress and self-esteem.* **Results:** *Women with MRKH had significantly more pathological scores on some of the scales and subscales, such as phobic anxiety and psychoticism (interpersonal alienation), with a similar trend for subscales measuring depression and anxiety.* **Conclusion:** *MRKH has a lasting negative impact on affected women's level of psychological distress and self-esteem.*

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Uterovaginal agenesis (Mayer-Rokitansky-Küster-Hauser Syndrome; MRKH) is a congenital abnormality of the female genital tract, resulting in nonformation of the vagina and the uterus, but with normal ovaries.[1] The etiology is speculated to be polygenic/multifactorial; occasionally the syndrome results from a genetic mutation or deletion of genes on Chromosome 16, but there remains no specific scientific explanation. Therefore, genetic counseling and pedigree analysis of family members of affected patients is of no value until such time as the human genome project is complete, the genes responsible for the genital tract are discovered, and a full understanding of the genetic basis of this condition is derived. Currently, we consider the condition to arise *de novo*.[2]

MRKH occurs in approximately 1 in 5,000 female births, and it is typically diagnosed in mid-adolescence.[1–3] Most of the literature on MRKH has focused on medical or surgical treatment outcomes for creation of a neovagina.[4,5] The treatment of choice for this is dilation therapy.[4] Success rates in creating a neovagina by use of the dilator approach is 100% in specialty centers.[5] A high proportion of women with MRKH do enter long-term relationships, and one study found that relationship satisfaction for these women is no different from that of control-group women.[5] With the advancement of reproductive techniques, having children of their own has now become possible for women with MRKH through in-vitro fertilization using a "carrier mother."[6] Importantly, no congenital abnormalities (of the uterus or vagina) have been found in children born by this technique.[1]

Relatively little is known about the psychological impact of MRKH. We recently conducted a systematic review on this topic.[7] Most of the articles identified were

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single case studies or small- to-medium-sized retrospective case series of women with MRKH who were followed up over a period of time. On the basis of the limited available evidence, the review tentatively concluded that adjusting to the diagnosis of MRKH is a difficult and traumatic process for these women, leading them to question their identity as women and to experience a sense of confusion regarding their gender, their bodies, and their social and sexual roles. This threat gives rise to the development of negative self-beliefs, with many women seeing themselves as defective, inferior, or unloveable. Surgical or dilator treatments are often experienced as shameful and may serve to perpetuate or strengthen these beliefs. Although the successful creation of a neovagina ameliorates some of these difficulties, there is general consensus that MRKH has a lasting effect, perpetuating these women's negative view of themselves. Their infertility, in particular, may serve to perpetuate these women's defective sense of self. As yet, no studies have formally investigated psychological functioning of women with MRKH and compared it to that of other women in terms of levels of psychiatric symptoms, interpersonal problems, self-esteem, or other aspects of psychological functioning.

The main aim of the present study was to conduct a cross-sectional comparison of the psychological characteristics of women with MRKH, as compared with a group of women without MRKH. We hypothesized that, compared with control-group women, those with MRKH would have significantly poorer functioning on a range of widely-used psychological self-rating scales. A second aim was to assess whether, over time, with increasing number of years since diagnosis, the psychological impact of MRKH would decrease.

METHOD

Subjects

Women on the MRKH register at the U.K. National Centre for Adolescent and Adult Women with Congenital Abnormalities of the Genital Tract were contacted by mail with information about a randomized, controlled trial (RCT) of group cognitive-behavioral therapy and, at the same time, were sent the set of psychological questionnaires listed here. Women age 17 or over, with a diagnosis of MRKH made or confirmed at the Centre, were eligible for the study. Women were told that, irrespective of their decision to participate in the RCT, we would be interested in their questionnaire results. Of 335 women on the register, 214 did not respond or were not contactable. Four women were not eligible for participation because they were below age 16; 78 declined to take part in the trial, 27 of whom were willing to complete the questionnaires. Thirty-nine women with MRKH decided to take part in the trial, all of whom completed baseline questionnaires. Thus, in all, 66 MRKH women (20%) had usable questionnaires. Control-group women were recruited from a London City International Church congregation, and from the City University (London) student population. The women were given a brief explanation by the researcher of the purpose of the study. Women interested in participating were provided with an information sheet for the study, the study questionnaires, and a self-addressed, prepaid envelop. Participants completed their questionnaires anonymously, but basic sociodemographic information was collected. Of 42 control-group women who were approached, 31 (73.8%) returned completed questionnaires.

Table 1 gives the characteristics of the study participants, which were broadly comparable between groups, except that women with MRKH more often were in a relationship ($\chi^2=4.04$; df: 1; $p=0.05$), and there was also a significant between-group difference in terms of social class ($\chi^2=8.92$; df: 3; $p=0.05$).

Measures

A number of widely-used, well-validated self-report questionnaires were used. These included 1) The Symp-

TABLE 1. Sociodemographic Characteristics

Variable	MRKH Women	Control Subjects	p
Age, years	27.9 (1.0)	27.8 (1.5)	NS
Ethnicity			
White	83.3%	67.7%	NS
Other	16.7%	32.3%	
Relational status			
With partner	66.7%	45.2%	0.05[a]
No partner	33.3%	54.8%	
Children (including step or adopted)			
Yes	12.1%	16.1%	NS
No	87.9%	83.9%	
Social class			
High	24.2%	41.9%	0.05[b]
Medium-to-low	46.9%	16.1%	
Student	25.8%	35.5%	
Housewife	3.0%	6.4%	

MRKH: Mayer-Rokitansky-Küster-Hauser syndrome; SD: standard deviation.
[a] $\chi^2_{[1]}=4.04$
[b] $\chi^2_{[3]}=8.92$.

tom Checklist (SCL-90–R),[8] which assesses a wide range of psychopathology and psychological distress factors and has been used in many different patient groups, including medical outpatient populations; 2) The Rosenberg Self-Esteem Scale (RSE),[9] which measures overall feelings of self-worth or self-acceptance; 3) The Inventory of Interpersonal Problems (IIP–32),[10] which measures interpersonal distress; and 4) The Eating Disorder Inventory (EDI),[11] which was included because there is anecdotal evidence in the literature that eating-disorder symptoms may develop in response to being diagnosed with MRKH.

Statistical Analysis

Data were analyzed with SPSS Version 13. Two group comparisons between MRKH women and comparison women on questionnaire measures were conducted, using independent-sample *t*-tests (two-tailed). In the MRKH group, we used Pearson correlation to analyze the relationship between questionnaire scores and years since diagnosis.

RESULTS

The comparison of women with MRKH and control-group women on the questionnaire measures is shown in Table 2. On the SCL-90–R, MRKH women had significantly higher scores on the subscales Phobic Anxiety and Psychoticism (interpersonal alienation), with a similar trend for the subscales Depression ($p=0.089$) and Anxiety ($p=0.087$).

On the Rosenberg Self-Esteem scale, women with MRKH had significantly higher scores (i.e., lower self-esteem) than control-group women. There was no difference between groups on the IIP–32. Women with MRKH had significantly higher EDI total scores than comparison women. In contrast to comparison women, the MRKH group had significantly higher scores on the subscales Interoceptive Awareness, Interpersonal Distrust, Ineffectiveness, and Bulimia.

In the MRKH group, the mean number of years since diagnosis was 9.6 (standard deviation [SD]: 8.7), with a

TABLE 2. Cross-Sectional Comparison of Women With MRKH and Control-Group Women

Variable	MRKH Group (N=66)	Control Group (N=31)	*t*	p
SCL-90–R (GSI)	0.90 (0.1)	0.67 (0.1)	1.52	0.133
SCL-90–R subscale scores				
Somatization	0.74 (0.1)	0.59 (0.1)	0.94	0.348
Obsessive-Compulsive	1.09 (0.1)	0.86 (0.1)	1.23	0.222
Interpersonal Sensitivity	1.13 (0.1)	0.95 (0.2)	0.89	0.377
Depression	1.27 (0.1)	0.92 (0.1)	1.72	0.089
Anxiety	0.78 (0.1)	0.53 (0.1)	1.74	0.087
Hostility	0.68 (0.1)	0.52 (0.1)	0.99	0.325
Phobic Anxiety	0.52 (0.1)	0.23 (0.1)	2.36	0.020
Paranoia	0.80 (0.1)	0.68 (0.2)	0.66	0.511
Psychoticism	0.73 (0.1)	0.46 (0.1)	1.96	0.054
Rosenberg Self-Esteem Scale (RSE)	22.8 (0.7)	19.3 (0.9)	2.77	0.007
Inventory of Interpersonal Problems (IIP–32)	38.8 (2.4)	34.5 (3.0)	1.04	0.299
Eating Disorder Inventory (EDI total)	40.4 (4.2)	26.8 (3.7)	2.41	0.018
EDI subscale scores				
Drive for Thinness	3.5 (0.7)	4.2 (0.8)	−0.70	0.486
Interoceptive Awareness	4.7 (0.7)	1.8 (0.5)	3.43	0.001
Perfectionism	4.6 (0.5)	4.2 (0.7)	0.44	0.662
Interpersonal Distrust	4.0 (0.6)	2.3 (0.5)	2.25	0.027
Ineffectiveness	6.7 (1.0)	2.5 (0.7)	3.42	0.001
Body Dissatisfaction	10.7 (1.0)	8.2 (1.4)	1.41	0.161
Maturity Fears	4.5 (0.6)	3.2 (0.6)	1.48	0.143
Bulimia	1.6 (0.4)	0.5 (0.2)	2.43	0.017

Values are mean (standard error). GSI: Global Severity Index (computed by first summing the scores on the SCL's nine symptom dimensions and a number of additional items; the sum is then divided by the total number of responses).

MRKH: Mayer-Rokitansky-Küster-Hauser syndrome.

range from 1 to 39 years. In this group, a correlational analysis was performed for years-since-diagnosis and questionnaire scores, to see whether psychological distress lessened over time. The correlation between years since diagnosis and SCL-90–R was 0.014 ($p=0.914$), RSE: $r=0.068$ ($p=0.587$), IIP–32: $r=0.109$ ($p=0.384$), and EDI: $r=0.045$ ($p=0.717$), respectively. None of these correlations were significant.

DISCUSSION

Women with MRKH differed from control-group women of similar age and ethnicity in terms of some of the subscale scores on the SCL-90–R and EDI, on the EDI total score and on the RSE, in the direction of more pathological scores. Thus, our hypothesis was partially confirmed. The lack of difference between MRKH and comparison women on the IIP–32 was somewhat unexpected, but may reflect the fact that this instrument measures relatively broad aspects of interpersonal relationship patterns. On the whole, impairments in the MRKH group were subtle. On the SCL-90–R, the scores of MRKH women lie somewhere between population norms for nonpatient and psychiatric outpatient scores.[12] On the RSE, although MRKH women had significantly lower self-esteem than control-group women, the mean RSE scores for both groups were in the normal range (i.e., 15–25 points). Low self-esteem, for whatever reason, is a well-documented risk factor for eating pathology.[13] The EDI measures both eating pathology and some of the associated personality traits. Women with MRKH had significantly higher scores than comparison women on the EDI total score and several of the subscales. Three of these subscales related to self-efficacy and self-worth (i.e., the Ineffectiveness subscale), the ability to form close, trusting relationships (i.e., the Interpersonal Distrust subscale), and the inability to identify emotions and physical needs and sensations, such as hunger or satiety (i.e., the Interoceptive Awareness subscale). Only one of the subscales on which MRKH women had higher scores than control subjects, namely, the Bulimia subscale, relates to overt eating-disorder symptoms. This suggests that MRKH women may try to compensate for lowered self-esteem and interpersonal difficulties by developing eating pathology.

The lack of more dramatic differences between the groups may reflect selection bias, given that we only managed to include 20% of potentially eligible women in our study. It is possible that those with the most prominent psychological disturbance chose not to participate. Alternatively, the literature on MRKH suggests that the greatest distress occurs immediately after diagnosis,[7] whereas most of the participants in the present study had lived with the knowledge of having MRKH for an average of about 10 years.

Contrary to our prediction, we did not find any significant correlations between time-since-diagnosis and psychological variables. Because our sample only had women who were well past the period of risk for distress, there may not have been enough variability in time-since-diagnosis to detect a significant association between this variable and psychological distress or impairment.

The study has a number of limitations, which include its cross-sectional nature and small sample size. In some instances, a failure to find differences between groups may have been due to lack of power. Moreover, MRKH participants and control subjects, although comparable in terms of age and ethnicity, differed in terms of social class and proportion living with a partner. Being in a partnered relationship is known to be a protective factor against psychological distress, so the fact that MRKH women were more commonly in a relationship than control subjects may have reduced differences in distress between them and the control women. On the other hand, the between-group differences in social class may have served to inflate differences between MRKH and control-group women, given that those of lower social class have been found to have more psychological distress.[14]

We did not screen MRKH women or control subjects for overt psychiatric caseness, and do not know how this might have affected our results. Furthermore, because only 20% of potentially eligible women chose to participate, it is not clear how representative this sample was of women with MRKH, in general. We believe that the reason for the low response rate is most likely related to our method of recruitment, which involved women being contacted by letter, with only written reminders. A more individualized approach to recruitment, (e.g., via telephone, or face-to-face in the clinic) might have yielded better results.

This study also has some strengths. The study and comparison-group sample were well-matched on some sociodemographic characteristics, and the response rate of the comparison group was high. The instruments are widely-used questionnaires. Perhaps the main strength of this study is that it adds to the very limited knowledge of psychological functioning in women with MRKH.

All in all, our study shows that, even in the context of a specialty service, where highly skilled support is easily accessible, MRKH has some lasting negative impact on

affected women's level of psychological distress and self-esteem. Future studies on women with MRKH may wish to chart the psychological effects of this condition longitudinally and specifically focus on the area of self-beliefs and gender- and sex-role behaviors. Likewise, in clinical services for MRKH women, brief assessment of psychological distress/functioning, at the time of diagnosis and at regular intervals thereafter, appears indicated, so as to identify those particularly in need of psychological intervention.

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