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Clinical characteristics, psychological effects, quality of life, and coping strategies in Chinese patients with Mayer-Rokitansky-Kuster-Hauser syndrome

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Summary

Objective: To investigate the clinical phenotype, psychological effects, and coping strategies of patients with Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome in China. *Materials and Methods:* The authors enrolled 46 patients with MRKH syndrome and collected the data on clinical manifestations by physical examination, psychological impact, and mental functions by self-administered questionnaires, and coping strategies through the medical questionnaire (MCMQ). *Results:* Among 46 patients with MRKH, 34 (73.91%) were classified as type I and 12 (26.09%) as type II; patients in the type II group had renal abnormalities and/or skeletal deformities, 60.86% (28/46) of the patients had feeling of anxiety, fear, and upset, 52.17% (24/46) were troubled by the disease, and 56.52% (26/46) were frustrated and had depression. However, 67.39% (31/46) of the patients were satisfied with their social status and family relations as well as perception of surrounding people. The results from the survey on the patients' strategies of coping with the disease showed that the mean scores (and SDs) of the facing, avoidance, and yielding dimensions were 18.30 ± 3.74, 17.05 ± 2.52, and 11.30 ± 4.37, respectively. The psychological function scores were significantly correlated with the scores of yielding dimension ($r = 0.613$, $p = 0.004$). *Conclusion:* The health impact of MRKH syndrome includes not only its physiological function, but also social and psychological aspects. The Chinese patients with MRKH syndrome often use yielding or avoidance strategies to cope with the disease. A comprehensive care and supporting program including medical, psychological, and social/educational components should be developed for MRKH patients in China.

Key words: MRKH (Mayer-Rokitansky-Kuster-Hauser) syndrome; Clinical phenotype; Psychology; Coping strategy.

Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a pathological condition characterized by vaginal agenesis and aplasia of the uterus [1-4]. This condition often affects a woman's ability to menstruate, to engage in penile-vaginal intercourse, and to bear children; the latter is often associated with infertility [1-4]. In individuals with MRKH syndrome, their karyotype (46, XX) and secondary sexual characters are mostly normal [1-3]. Although its etiology and pathology are not fully understood, MRKH syndrome is suggested to be caused by the lack of development of the Müllerian ducts between the fifth and the sixth weeks of gestation [5]. Patients with MRKH syndrome may also have renal agenesis or ectopia, spinal, and other abnormalities [6-16]. The genetic and genomic deficiencies in those individuals may vary, including mutations of several genes such as WT1, PAX2, HOXA7-HOXA13, PBX1, and WNT4 [17-26]; these genes are commonly involved in early embryonic development.

There are numerous published reports on this syndrome, mostly focusing on the clinical manifestations and surgical treatment, especially about how best to create a neovagina in women with MRKH [27-40]. However, little has been known about the psychological impact of MRKH and quality of life outcomes for women with the condition [1, 14]. Clinical research results have indicated that surgical or non-surgical creation of a neovagina can improve patient's sexual function, but may not lead to a successful psychological outcome [1, 14, 41-43]. Therefore, a strong psychological support from gynecologist and other healthcare professionals, patient's partner, family and relatives, as well as community, is critical and often helpful in coping with this syndrome [1, 14, 44].

Although MRKH syndrome in China is as common as other countries, there is limited research on this disease from China [45-56]; there is no report on the aspects of psychological effects and coping style of patients from China. The present study was designed to investigate the clinical phenotype, psychological effects, and coping style of patients with MRKH syndrome in China. This retrospective study reviewed 46 patients with MRKH syndrome from the present clinic and collected data in their clinical manifestations, psychological impact, and mental functions, and their coping strategies towards the disease. Considering the cultural differences between Chinese patients and patients from other countries, the authors developed

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novel survey questionnaires that adapted from published methods [44, 57-60]. The present results indicated that the health impact of MRKH syndrome on a patient's quality of life included not only the physiological aspects, but also social and psychological aspects. Therefore, a comprehensive medical, psychological, and social/educational caring and supporting program should be developed for these patients in China. The present results also indicate that cultural differences should be carefully taken into account in developing intervention approach to treating and caring patients with MRKH syndrome.

Materials and Methods

In the present study, the authors enrolled consecutive 46 patients with a confirmed diagnosis of MRKH syndrome that were seen in this hospital. The study design and clinical protocol were reviewed and approved by the Ethics Review Committee of the Affiliated Ninth People's Hospital, Jiao-Tong University Medical College, Shanghai, China. Each of the participants provided written informed consent prior to participating in the study. The inclusion criteria were: 1) a confirmed diagnosis of MRKH in the present hospital, 2) no prior treatment (surgical or non-surgical), and 3) consented to participate in this study. Exclusion criteria were: 1) prior diagnosis of MRKH or treatment for the disease before visiting the present hospital, 2) history of mental diseases, chronic diseases, or injury, 3) known trauma or other neuro-psychological disorders, 4) refusal to consent, and 5) incomplete questionnaire or missing data.

For each patient, demographic data such as age, education, occupation, family history, history of chronic diseases, injury, neurological diseases, psychological disorder and psychiatric diseases, medication, and treatments were recorded. Routine physical exam and laboratory test were performed, including chromosome analysis, ultrasound, spinal X-ray, female hormone, and MRI.

Self-report questionnaires were admitted to each of the participants before their surgery and after they provided informed consent. The questionnaire included two parts: Part I included 20 questions in the standard medical coping modes questionnaires (MCMQ) and Part II included 15 questions in the quality of life (QOL-BREF) questionnaires. During the development and validation of the MCMQ and QOL-VREF questionnaires, the authors referred to the published instruments and procedures and modified them accordingly [57-60]. The MCMQ questions included three dimensions, *i.e.,* facing, avoidance, and yielding approaches. For each question, the participant chose one of four answers provided, corresponding to a score from 1 to 4. Among the 20 questions, eight were negatively phrased and therefore the scores were reversed to calculate the final score of the MCMQ. Each QOL-BREE question had five choices, corresponding to scores 1-5.

The Statistics Package SPSS13.0 was used to establish the dataset. The scores were expressed as mean and SDs. The differences were analyzed by Student's *t*-test or ANOVA. Spearman correlation analysis was used to determine the correlation between the scores of various dimensions in MCMQ, *i.e.,* facing, avoidance, and yielding approaches, and psychological function scores. $P < 0.05$ was considered statistically significant.

Table 1. — *Psychological impact and social support scores among 46 patients with MRKH.*

Eight psychological parameters	Answers (n)				
	No	Little	Yes	Some	Extreme
Are you happy?	10	10	24	2	0
Can you concentrate on your work?	3	8	9	24	2
Are you self–satisfied?	10	16	15	5	0
Do you feel happy with your appearance?	5	8	19	10	4
Do you have negative feelings (bad, hopeless, anxious, fear, or depressed)	1	5	19	10	11
Social support					
Are you happy with your social networking?	5	7	14	11	9
Are you happy with help/ support from your friends?	4	3	9	14	16
Are you happy with your sex life?	46	0	0	0	0

Results

The authors enrolled 46 patients into the present study. Among them, the female genital development appeared to be normal and other secondary female sex characteristics were also normal. They were classified into two types, according to the results of clinical physical exams. Thirty-four cases were classified as type I MRKH that was characterized by vaginal agenesis and aplasia of the uterus and 12 cases were type II that were patients with MRKH syndrome and also had renal agenesis or ectopia. Forty-five out of the 46 patients with MRKH syndrome had karyotype (46, XX) and one case had 46XY. None of the 46 MRKH cases had family history of MRKH. One case of MRKH had a twin sister with normal development.

Table 1 shows the answers of the participants to the main questions on psychological and mental functions and social supports. 86.95% (40/46) of the patients expressed unhappiness with their life, 60.86% (28/46) often had feeling of anxiety, fear, and upset, 52.17% (24/46) were troubled by the disease, and 56.52% (26/46) were frustrated and had depression. However, 67.39% (31/46) of the patients expressed their satisfaction with their own social and family status, as well as perception of surrounding people towards their disease and social functions. All the patients were not satisfied with their sex life.

The results of the survey on the patients' strategies for coping with the disease showed that the mean scores (mean ± SDs) of the facing, avoidance, and yielding dimensions were 18.30 ± 3.74, 17.05 ± 2.52, and 11.30 ± 4.37, respectively. The psychological function scores were significantly correlated with the scores of yielding dimension ($r = 0.613$, $p = 0.004$). There was a trend in the correlation between

psychological function scores and facing and avoidance dimensions, but not statistically significant ($r = 0.379$, $p = 0.099$; $r = 0.389$, $p = 0.090$).

Discussion

In gynecological practice, MRKH syndrome is an uncommon, but not rare, congenital anomaly of the female genital tract [1-5, 13, 14, 44-46]. It is the second most frequent cause of amenorrhea after gonadal dysgenesis and is often discovered when a patient presents in adolescence due to primary amenorrhea [1-5]. MRKH syndrome is one of disorders of sex development (DSD) that refers to congenital conditions in abnormal development of chromosomal, gonadal, or anatomic sex. As a DSD, MRKH syndrome poses challenges that go far beyond physical concerns [1-5, 44]. A young woman's sense of well-being and quality of life are significantly impacted by the condition for long time, from diagnosis, to therapy, to daily life. Affected individuals without treatment will find it impossible or difficult to engage in penile-vaginal intercourse, do not menstruate, and will be unable to carry a pregnancy. The discovery that sexual intercourse will not occur without medical intervention and the realization of lack of childbearing ability may be devastating to an adolescent who has not yet reached certain developmental milestones and even has limited understanding of medical terminology.

To cope with physical and psychological challenges, the treatment of MRKH not only needs to anatomically manage the anomaly of the female genital tract so that young women may have the option to engage more easily in penile-vaginal intercourse, but also needs to help young women cope with the psychological and social impact of this syndrome [1-5, 13, 14]. Thus far, most clinical researches have emphasized the outcomes of surgical and non-surgical treatments to create a new vagina, there have been limited reports investigating the psychological impact and QOL of individuals with the syndrome [44]. Additionally, it has been recommended that a shift in emphasis takes place from the physical aspects of MRKH syndrome to how individuals adjust to the conditions [1-5, 13, 14]. The results from the present study support this shift. The present authors believe that psychological issues, mental function, and coping strategies should be addressed in a comprehensive manner in order to provide optimal care to the individuals with MRKH syndrome.

The QOL issue is one of the most important aspects in management of MRKH syndrome [44]. It has been suggested that QOL studies should take into account a person's physical, psychological, social, and spiritual dimensions to deal with a complex health problem such as MRKH syndrome. Having MRKH syndrome affects a woman's QOL by placing limits on some of life's capabilities, such as intercourse and childbearing, resulting in distress and an altered self-concept in patient's daily life, as well as relations with other people (most relatives, co-workers, and people who have knowledge of her disease status). In the present study, the authors paid close attention to how women were functioning in regards to specific QOL domains and how the women were impacted psychologically due to MRKH syndrome. The present results were organized according to these dimensions of QOL and psychological effects and coping methods.

Several studies have indicated that cultural tradition towards marriage and childbearing plays a major role in patient's own attitude to coping with MRKH syndrome and the attitude of her surrounding people, such as perception of the disease and the social role and status of the affected individual [44]. It is generally more difficult to accept and cope with the MRKH syndrome for women from more traditional cultures, where childbearing is expected and sometimes necessary to survival. China is among the traditional cultures. For example, a man from Bangladesh may abandon a woman with a condition such as MRKH [61]. An India woman with MRKH may not seek medical help until the time of marriage arrangement; the family may consider marrying the daughter to a widow who has already completed his family or to a man who is physically challenged and wants to adopt children [62]. This is almost true for Chinese culture, indicating that traditional cultural factors should be considered as an integral part of the intervention approach when the impact of MRKH on a woman's QOL is dealt with.

Psychological effects of diagnosis of MRKH on the affected individuals have a wide range of reactions, mostly negative, as reported in the present study. These responses included reactive depression, shock, feeling different, and confused. In addition, fear of rejection and isolation were common. In some of the women, a diagnosis of MRKH was likened to have a trauma, leading to persistent psychological distress. Individual adjustment to MRKH may vary significantly among the affected women. In the present study, the majority of women with MRKH adjusted to the condition reasonably well, making a positive adjustment to the disorder. The present authors found that 67.39% of the patients were generally well-adjusted and happy with their appearance and their social networking. Women with MRKH used a variety of strategies to cope with this diagnosis. Coping strategies ranged from denial to compensation by accomplishment. Working towards life goals was found to be a positive coping technique. In the present study, the patients reported that they could concentrate on their work and daily life after the diagnosis of MRKH. Families varied in their responses to their daughters' diagnoses. Parental reactions were reported to range from guilty, difficulty adjusting to gentle and supportive. Parental support was helpful to a positive outcome and it was recommended that parents be a part of counseling and decision-making. In the present study, the patients reported that their families and friends were generally supportive.

Many questions remain as how to best treat women with MRKH. The present study suggests that psychological counseling is urgently needed, especially for several critical time periods, such as diagnosis, treatment for a neovagina, relationships with men, and creation of a family. The strategies to cope with MRKH may vary and the affected individuals should be encouraged to. If to determine what are more operationally in dealing with MRKH and these coping techniques should be incorporated into treatment plans and supportive interventions for women with MRKH. Additionally, a women's level of knowledge about her MRKH condition may affect her ability to cope with it. Therefore, during counseling and treatment, it is important to evaluate the woman's knowledge of MRKH and how it contributes to her own healthcare self-efficacy.

Finally, information management is an important issue in managing MRKH, including informing patients about their diagnosis and sharing information with family members and the wider community. In China, talking about a condition that involves the genitals and sexuality can be difficult for many people, including adolescents and adults. Future study should investigate the approaches to information sharing such as to whom, when, how to best disclose or share information about the condition and how much information is useful to patients and appropriate for others to know.

Of note, there were some limitations of the present study. First, all the patents were not treated for their physiological abnormalities. There are many studies suggesting that, after surgical treatment to create a neovagina, many women with MRKH syndrome are able to engage successfully in penile-vaginal intercourse, which may have a significant impact on the patients' social and psychological performance [1-5, 13, 14]. Future studies should investigate the status and QOL of Chinese women with MRKH syndrome who undergo surgical treatment. Second, the present study did not address the effects of MRKH syndrome on marriage and cohabitation. The present authors did not know if our patients were married nor had a steady sexual partner. Literature review suggests that marriage or steady sexual partnership would affect the QOL of patients with MRKH syndrome [1-5, 44].

Based on the present results, the authors speculate that there is a need for better research and management for Chinese women with MRKH syndrome. Optimal care for women with MRKH includes both medical and psychological support at important milestones throughout a woman's life span. A medical approach to treating MRKH is necessary, but not sufficient [1-5, 13, 14, 44]. Physicians can further assist young women by being very thoughtful and deliberate in the manner that they reveal the diagnosis of MRKH. Physicians can also provide information about the condition and provide referrals to mental health professionals and to support groups. Screening newly diagnosed patients for degree of psychological distress can help identify those individuals who may benefit from longer-term counseling. Further investigation into the factors that mediate and/or moderate the impact of MRKH will assist healthcare professionals in improving medical and psychological care for women with MRKH.

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