

A Cognitive Behavioural Model and Therapy for Utero-Vaginal Agenesis (Mayer-Rokitansky-Küster-Hauser Syndrome: MRKH)

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Background: Utero-vaginal agenesis, also called the Mayer-Rokitansky-Küster-Hauser Syndrome (MRKH), is a congenital abnormality of the female genital tract, characterized by the non-formation of the vagina and the uterus. It is a common cause of primary amenorrhoea. Little is known about the psychological impact and management of this condition. **Method:** We describe a specific model of the core negative psychological impact of diagnosis and medical treatment of MRKH and a cognitive-behavioural therapy of MRKH based on the model (CBT-MRKH). The Medical Research Council's (2002) framework for the development and evaluation of complex health interventions was used for intervention development and evaluation. **Results:** Evidence from a recent cross-sectional study and a small randomized controlled trial (RCT) provides preliminary support for the model and treatment (Heller-Boersma, Schmidt and Edmonds, in press; Heller-Boersma, Schmidt and Edmonds, 2007), and this is further validated by extensive qualitative material gathered over the course of the RCT from participants. **Conclusions:** The model and treatment described may also be applicable to a number of other congenital or acquired gynaecological conditions such as premature ovarian failure, breast cancer, early onset endometrial cancer, female genital mutilation, Turner's Syndrome, ovarian dys/agenesis or, Complete Androgen Insensitivity Syndrome, all of which have a psychological impact not dissimilar to MRKH in terms of these women's sense of self and femininity.

Keywords: Utero-vaginal agenesis, Mayer-Rokitansky-Küster-Hauser Syndrome, cognitive-behavioural therapy, psychological management of MRKH, adjustment to diagnosis and treatment of MRKH.

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Introduction

Uterovaginal Agenesis, also called the Mayer-Rokitansky-Küster-Hauser Syndrome (MRKH), is a congenital abnormality of the female genital tract. It is characterized by the non-formation of the vagina and the uterus, but with normal ovaries and normal secondary sexual characteristics. A proportion of affected women have other abnormalities (kidney, ear or skeletal) (Strubbe, Cremer and Willemsen, 1994). In the majority of cases the aetiology is multifactorial (Edmonds, 2003). MRKH occurs in about 1 in 5000 female births (Aittomaki, Eroila and Kajanoja, 2001). It is the second most common cause of primary amenorrhoea after Turner's Syndrome (Edmonds, 2000). Affected women are typically diagnosed in mid-adolescence after failure to menstruate (Edmonds, 2003). In the past, surgical approaches to the creation of a neovagina were favoured, whereas the current treatment of choice is dilation therapy (Frank, 1983; Nadarajah, Quek, Rose and Edmonds, 2005; Ismail-Pratt, Bikoo, Liao, Conway and Creighton, 2007). This involves the patient being taught to put pressure on the vaginal dimple using dilators of gradually increasing size for 20 minutes three times a day over several months (Edmonds, 2000). Women with MRKH are infertile and having children of their own is only possible through in vitro fertilization using surrogacy (e.g. Beski, Gorgy, Venkat, Craft and Edmonds, 2000).

Clinicians treating this condition agree that it has a significant psychological impact, yet little is known about the precise nature of any psychological sequelae and how this might be translated into an intervention. The aims of this paper are: 1) to describe the development and content of a specific model of the core negative psychological impact of diagnosis and medical treatment of MRKH and how this is maintained; 2) to describe the development, structure and procedures of a cognitive-behavioural therapy of MRKH based on the model (CBT-MRKH); and 3) to present preliminary evidence that supports the model and treatment.

Development of a model of the psychological impact of MRKH

The work presented here was based on the Medical Research Council's (2002) framework for the development and evaluation of complex health interventions. We initially conducted a systematic review of the MRKH literature (Heller-Boersma, Edmonds and Schmidt, submitted) focusing on the psychological sequelae of the diagnosis of MRKH and its treatment, so as to develop ideas about targets for intervention. The review was limited by the poor methodological quality of many of the available papers. Out of 479 identified sources, only 16 were case reports or series ($n = 1$ to 79) with a primary focus on psychological, psychosocial or psychosexual aspects of MRKH. Seven further case series had a mainly medical or surgical focus but with some mention of psychological aspects of MRKH.¹ Only one series was prospectively gathered and only one study included a comparison group of healthy women. Findings were mainly descriptive and often presented without fully quantifying them (e.g. by saying "most patients" or "many patients"), thereby making it hard to distinguish between authors' impressions and data obtained from the patients. Nonetheless, several key themes emerged from the review. These are briefly described below:

¹A full list of references included in the systematic review can be obtained from the authors upon request.

(a) Immediately following diagnosis patients report a broad range of emotional responses from positive (including relief, fighting back) through to more negative responses such as shock, confusion, fear, depression and suicidality, feeling isolated, ashamed, or guilty through to more avoidant/cut-off responses such as dismissal or denial). Several authors noted that despite being given careful and detailed descriptions of their condition and its treatment, many young women were not fully able to retain the information and had major misconceptions about this.

(b) Being given the diagnosis of MRKH and the implications this has for their sexual and reproductive lives creates a major threat to these young women's sense of themselves, leading them to question their identity as a woman and to experience a sense of confusion regarding their gender, bodies, 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. Whilst the successful creation of a neovagina ameliorates some of these difficulties, MRKH often seems to have a lasting negative impact on these women's view of themselves. In particular, the infertility associated with MRKH may serve to perpetuate these women's defective sense of self.

(c) Many women seem to try to control the sense of threat to their femininity and the accompanying symptoms by using potentially maladaptive coping strategies.

(d) There is widespread agreement in the MRKH literature that psychological support or intervention for these women is important, yet there is little detail on what such an intervention might look like.

Overview over the model

Based on our review of the MRKH literature, our clinical experience and adapted from the CBT-model of PTSD by Ehlers and Clark (2000) and Ehlers, Clark, Hackmann, McManus and Fennell (2005) we postulated three key mechanisms that lead to and serve to maintain a problematic adjustment to MRKH. These are: (a) a failure to "process" the diagnosis; (b) the occurrence of idiosyncratic negative appraisals of MRKH-related events that become incorporated into these women's core schemata and sense of identity as women; (c) women's attempts to control the sense of threat to their femininity and the accompanying emotional symptoms by using a range of potentially maladaptive cognitive and behavioural strategies. These processes occur in a clear developmental context, a point that is almost entirely missing from the literature on MRKH. Below, we will give details first on the developmental context, then on the processes underpinning poor adjustment to MRKH.

The developmental context of MRKH related "events"

Most of the MRKH-related "events" (diagnosis, treatment) unfold during a narrow developmental window from early into late adolescence. The onset of puberty is considered as one of the most important developmental milestones for girls to negotiate (Brooks-Gunn and Reiter, 1990; Stattin and Magnusson, 1990). In particular the first menstruation is considered by many to be the "single most critical event marking puberty in girls, signifying and symbolizing feminine identity and maturity, attributes that will constitute the core of the personality in subsequent development towards adult status" (Stattin and Magnusson, 1990). Adolescents'

perception that they are developing “on time” or are later maturing than their peers, may be as important or even more important than actual timing of puberty and related milestones (Dubas, Graber and Peterson, 1991). In the UK, the median age at menarche is now about 13 (Whincup, Gilg, Odoki, Taylor and Cook, 2001), i.e. 3 or 4 years earlier than the typical age of diagnosis of MRKH. Thus even prior to diagnosis young women with MRKH may perceive themselves as seriously “out of sync” with their peers.

The reported mean or median age at diagnosis of MRKH is between 16.0 to 17.7 years (Heller-Boersma et al., 2007). This is a time when these young women are still somewhat cognitively immature (Haffner, 1995), i.e. may have a tendency to rule-based black-and-white thinking, which is likely to foster poor processing of MRKH related events and the development of dysfunctional beliefs and coping strategies.

Moreover, MRKH related events happen during the time when in most Western countries these young women typically are expected to take important sexual milestones of development. For example, in various Western samples, the median age at first date was 14.4 years, the median age at first petting was 15.8, and the median age at first intercourse was about 17 years (ACSF, 1992; Schalet, 2004; Schmidt, Evans, Tiller and Treasure, 1995).

Processes underpinning poor adjustment to MRKH

Failure to “process” the diagnosis. It is known from work in cancer that people’s memory for “bad news consultations” is often fragmented and not organized in a coherent narrative, with misconceptions about what was said and what is wrong (Fallowfield and Jenkins, 2004). This is on a continuum with what happens in the development of Post-Traumatic Stress Disorder, where trauma memory is poorly elaborated and not incorporated fully into the autobiographical memory base (Ehlers and Clark, 2000). The literature on MRKH suggests that similar processes may be at work here. For example, one of the early authors in the MRKH field notes that, “the reaction to diagnosis is replete with painful confusion and shocked misunderstandings” (Kaplan, 1968). In our clinical experience, when patients are asked to describe how they were diagnosed with MRKH, they often remember and focus on what seems peripheral detail. For example, they may describe the dress they wore on that day, its color and pattern, what the weather was like, or remember details about the consultant’s appearance or their office. However, there typically are also gaps in the story, where they cannot remember what happened. These anecdotal observations point towards a re-experiencing of being given the diagnosis mainly in terms of sensory impressions rather than in the form of a coherent narrative, possibly as a result of the information being too overwhelming and painful, and associated with poor intentional recall (Ehlers and Clark, 2000). This makes it hard for the person to make sense of what happened, and prevents the updating, revising and incorporation of new information and therefore the ability to adjust to the MRKH diagnosis.

The development of idiosyncratic negative appraisals of MRKH-related events. Having congenital agenesis of the uterus and the vagina, perhaps combined with other congenital abnormalities e.g. of the kidneys, would be very distressing to anybody. Appraisals such as “I am different”, “life is not fair”, “my prospects for finding a partner are reduced” are to some extent realistic. Nonetheless, there will be many appraisals related to the events of MRKH that are overly negative and idiosyncratic, due to the cognitive immaturity, lack of experience and rule based black-and-white thinking of these young women. Table 1 shows some such

Table 1. Developmental context of MRKH related events and difficulties and associated negative appraisals

Developmental context	What is appraised	Negative appraisal
<p><i>Early to mid adolescence (age 12 to 16):</i></p> <p>Cognitive development: concrete thinking</p> <p>Girlfriends start to menstruate (mean age menarche age 12)</p> <p>Importance of being like peers</p> <p>Increasing importance of body image</p> <p>Early sexual experimentation, e.g. first kiss, first date</p>	<p>Failure to menstruate at the expected time</p>	<p>“I am different to other girls”</p> <p>“I am not a proper woman”</p>
<p><i>Mid to late adolescence (age 16 to 17)</i></p> <p>Increased abstract thinking ability</p> <p>Experimentation with relationships and sexual relationships</p> <p>First petting; first sexual intercourse</p>	<p>Being diagnosed:</p> <p>Being told you have no vagina and uterus</p> <p>Being told you are infertile</p> <p>Doctor’s behavior during consultations: e.g. focusing on technical and medical aspects of treatment</p> <p>Parents’ response to diagnosis: e.g. depression, denial, shame</p>	<p>“I am not normal”</p> <p>“I am not a proper woman”</p> <p>“I am sexually neuter”</p> <p>“I am incomplete/a freak/soiled goods”</p> <p>“I am worthless as a woman”</p> <p>“If this can happen, anything can happen, the world is a scary place”.</p> <p>“I am a freak”</p> <p>“I am not a proper person”</p> <p>“I am outside of female experiences</p> <p>“There is no place for me in society – the future is bleak”</p> <p>“I am unloveable”</p> <p>“I must never mention this”</p> <p>“I am an embarrassment to others”</p> <p>“I am all alone”</p>
<p><i>Late adolescence (age 17–19):</i></p> <p>Cognitive and emotional development nearly secured</p> <p>Body image and gender role identification nearly secured</p> <p>Sexual orientation nearly secured Greater intimacy skills Transition to adult roles</p>	<p>Pain/discomfort related to gynaecological interventions: e.g. having to use dilators</p>	<p>“Sex is disgusting”</p> <p>“I am disgusting”</p> <p>“I deserve this”</p>

Table 1. Continued.

<i>Any age thereafter</i>	Being confronted with other women's sexuality: e.g. hearing friends talk about periods, being asked for a tampon, seeing other women in relationships, or with their children or having a surrogate mother carrying one's child	"I am bad for feeling this way (envious or left out)" "I am inferior" "I am defective, incompetent, useless, inadequate"
	Relationship with boy-friend/partner: e.g. assumed or actual rejection	"Nobody will ever want me" "I am unacceptable as a sexual partner" "I am unloveable" "Men will always dump me" "Men can't be trusted" "I am second best – can only get second best"
	Further medical or gynaecological procedures (e.g. in vitro fertilization)	"I am defective"

appraisals in their developmental context, together with the events that might elicit them. The response of others may inadvertently strengthen and reinforce such appraisals.

So far we have outlined the development of a problematic adjustment to MRKH during adolescence arising directly from the MRKH-related events as they unfold. However, in the longer term any idiosyncratic negative appraisals of MRKH-related events are likely to become incorporated into these women's core schemata concerning their body and their sense of identity as women.

For many young women with MRKH, their body schema, i.e. a woman's internal mental representation of her body on which – according to Piaget (1952) – all other schemata are built, is shattered. Being given a diagnosis of an "absence of a vagina and uterus" is hard to make sense of. Incoming stimuli that confront her can no longer be coded and matched with what she previously knew about her body and its integrity. Learning about other abnormalities that occur in the context of MRKH, such as renal abnormalities, may further compound the threat to the body schema and further alienate her from her body. In parallel, the young woman's schema of herself as a developing woman who can have sexual and love relationships and children is shattered, with the future seeming bleak or at least very uncertain.

To this a new MRKH-related schema is added. We say this because many of our patients talk about how the MRKH becomes their major preoccupation and identity, through which all other experiences are filtered and coded. The body, feminine and MRKH related schemata, which are closely linked, become imbued with a negative meaning of being different, defective, sexually inadequate, worthless, unloveable, out of control or even outside human experience. The exact nature and cognitive/emotional colouring of this is likely to depend on a number of factors such as other pre-existing experiences and/or schemata. One powerful contributing factor is the meshing of the view of self with culturally derived beliefs about what it means to

be a woman, where self-worth or value becomes narrowly linked to the presence of a vagina, the ability to have sex, to have a womb and give birth to children.

Use of potentially maladaptive cognitive and behavioural strategies for the reduction of threat. Women are likely to attempt to control the sense of threat to body and their femininity and the accompanying emotional symptoms by using a range of potentially maladaptive cognitive and behavioural strategies. The strategies chosen are likely to be meaningfully linked with the individual's appraisals of a particular situation (Ehlers and Clark, 2000). For example, a young woman with a core belief of "I am inadequate and defective as a woman" may develop a rule such as "I will only be acceptable to people if I present as the perfect woman". This might make her spend huge efforts to try to improve her appearance and make herself desirable as a woman, in terms of dress, make-up or a perfect figure. At times when this becomes too exhausting she may flip into alternative behaviours, abandoning her striving for perfection and she may binge eat, generally neglect her appearance and abandon efforts to appear feminine.

Alternatively, women may act in keeping with their beliefs about themselves. An example would be a woman with MRKH who chooses partners who she recognizes are unsuitable and allows herself to be used and treated badly by them because she believes "No one else would want me, I am unacceptable and unlovable". This woman might react with suspicion when a good man shows interest in her and so reject him because she believes "there must be something wrong with him if he shows interest in someone as defective as me".

These strategies intended to reduce the threat of MRKH are maladaptive because they maintain the threat associated with MRKH by three mechanisms: (1) directly producing psychological symptoms; (2) preventing change in negative appraisals of MRKH or its sequelae; and (3) preventing change in the processing of the diagnosis (cf. Ehlers and Clark, 2000). Table 2 shows how idiosyncratic beliefs may relate to cognitive and behavioural strategies and the effects this has on the person.

Additional considerations

Appraisals and emotional responses. Adjustment reactions to bad news are typically characterized by intense and mixed emotions, such as anger, fear, and depression, often rapidly alternating (Moorey, 1996). Particular emotions are meaningfully linked to particular appraisals (Ehlers and Clark, 2000). One salient feature of MRKH is that many women constantly compare themselves (negatively) against other women and this is associated with appraisals such as "I am inferior", "I am not a proper woman". Emotions aroused by situations involving social comparison where there is a threat to rank prominently include shame, guilt, envy or jealousy (Gilbert, 2001). These emotions are generally seen as socially unacceptable and people tend to keep such emotions to themselves. They also have the potential for contributing to and intensifying a person's sense of themselves as a bad person and as harbouring something that needs to remain hidden.

Triggers

Schema-level beliefs and associated rules or assumptions can lie dormant but become activated by salient triggers. In the case of MRKH, triggers or critical events often are related to

Table 2. Idiosyncratic beliefs and their relationship to cognitive and behavioural strategies used to compensate and the effects this has on the person

Idiosyncratic appraisals	Cognitive or behavioural strategies	Effects on the person
<i>In relation to self (body, femininity and sexuality):</i>		
e.g. I am defective I am not a proper woman I am worthless as a woman I am a freak of nature I am unloveable	a) Trying to compensate by: being the best in other areas (e.g. work or study), or by focusing on having the perfect feminine appearance, weight an shape, or by always fitting in or by pleasing others b) Avoid difficult MRKH related thoughts and feelings by: alcohol, drugs, self-starvation or binge eating	a) Own needs ignored → exhausted, angry, resentful, depressed b) Substance misuse or eating related symptoms → feel worse about self
<i>In relation to others (partners, family):</i>		
e.g. Nobody will want me Men will always dump me Sex is disgusting Men can't be trusted I am an embarrassment to my family	a) Try to compensate by: being promiscuous b) Settle for "second best" in relationships c) Avoid intimate relationships d) Keep MRKH a secret	a) Increased risk of sexually transmitted disease, increased risk of being treated badly by casual partners b) Further undermines self-esteem, no opportunity for disconfirmation c) Forgo opportunity for loving close relationship d) Feel miserable and alone, no opportunity for disconfirmation
<i>In relation to the world and future:</i>		
Life is not fair The world is an unpredictable and unsafe place	a) Dwell on "why me? It is not fair" b) Try to be in perfect control of life, plan everything 100%, leave nothing to chance	a) Keeps person stuck and depressed b) Exhausted

dating/courtship or to relationship break-ups, which bring down a carefully built up "façade" and activate the woman's negative beliefs about herself. Alternatively, events such as a friend or a sister having a baby may also serve to activate MRKH-related schemata, as may the medical procedures accompanying in vitro fertilization using surrogacy. Overtime, an increasingly greater range of triggers that are only peripherally or not at all related to MRKH may come to elicit women's beliefs and compensatory strategies.

Differences between poor adjustment to MRKH and PTSD

There is a key difference between what happens in people with a problematic adjustment to MRKH and what happens in PTSD. In PTSD the sense of current severe threat is, above all, maintained psychologically through the nature of the trauma memory, the resulting negative appraisals, and the strategies intended to control threat or symptoms (Ehlers and Clark, 2000). In contrast, in MRKH, in addition to these processes it is the unalterable presence of the actual physical impairment that underpins and feeds into the sense of current threat.

Developing a CBT treatment of the psychological sequelae of MRKH (CBT-MRKH)

Several potential targets for psychological intervention follow from our model: First, treatment should revisit women's experience of being diagnosed and the events surrounding this with the aim of helping them to fully process what has happened and integrate it "*into the context of the individual's preceding and subsequent experience...*" (Ehlers and Clark, 2000). Second, treatment should help women to examine their problematic self-appraisals and beliefs that have arisen in the context of their MRKH treatment and relationship history and help them to challenge and modify these as they maintain the current sense of threat to their sense of themselves (see Ehlers and Clark, 2000). In particular, these women need to be helped to develop a more positive view of their own femininity. This may mean helping women to shift from a narrow, caricatured view of femininity as derived from the possession of a vagina and womb to a broader, more balanced and more accepting view of what makes a woman loveable, acceptable and competent. Third, they need to develop a more positive, accepting view of their MRKH as something that is no longer a shameful secret, but something that can be thought and talked about and lived with. This work towards greater acceptance of things that can not be changed needs to include an acknowledgement and mourning of the losses incurred as a result of MRKH. Finally, treatment should address any maladaptive compensatory or coping strategies that may serve to maintain unhelpful self-beliefs.

Structure of CBT-MRKH

We decided that a group intervention would be preferable to individual therapy, as this would help women to challenge the commonly held belief that they are "abnormal" or "a freak". The idea of a group intervention is also supported by the MRKH literature, which highlights the importance of women having the chance to communicate with others with this diagnosis (for review see Heller-Boersma et al., submitted).

Overview of contents of CBT-MRKH

A formulation-based approach. The intervention centres around a longitudinal cognitive-behavioural case conceptualization (Beck, 1996; Padesky; 1994) that serves as an overall working model, and is used to explain the CBT model and to guide the therapy. Each woman produces her own formulation, tailored to her particular circumstances. Figure 1 shows an example conceptualization diagram.

Re-processing diagnosis. Treatment starts with a focus on the central life event of being diagnosed, and aims to help women move from poorly processed patchy memories and

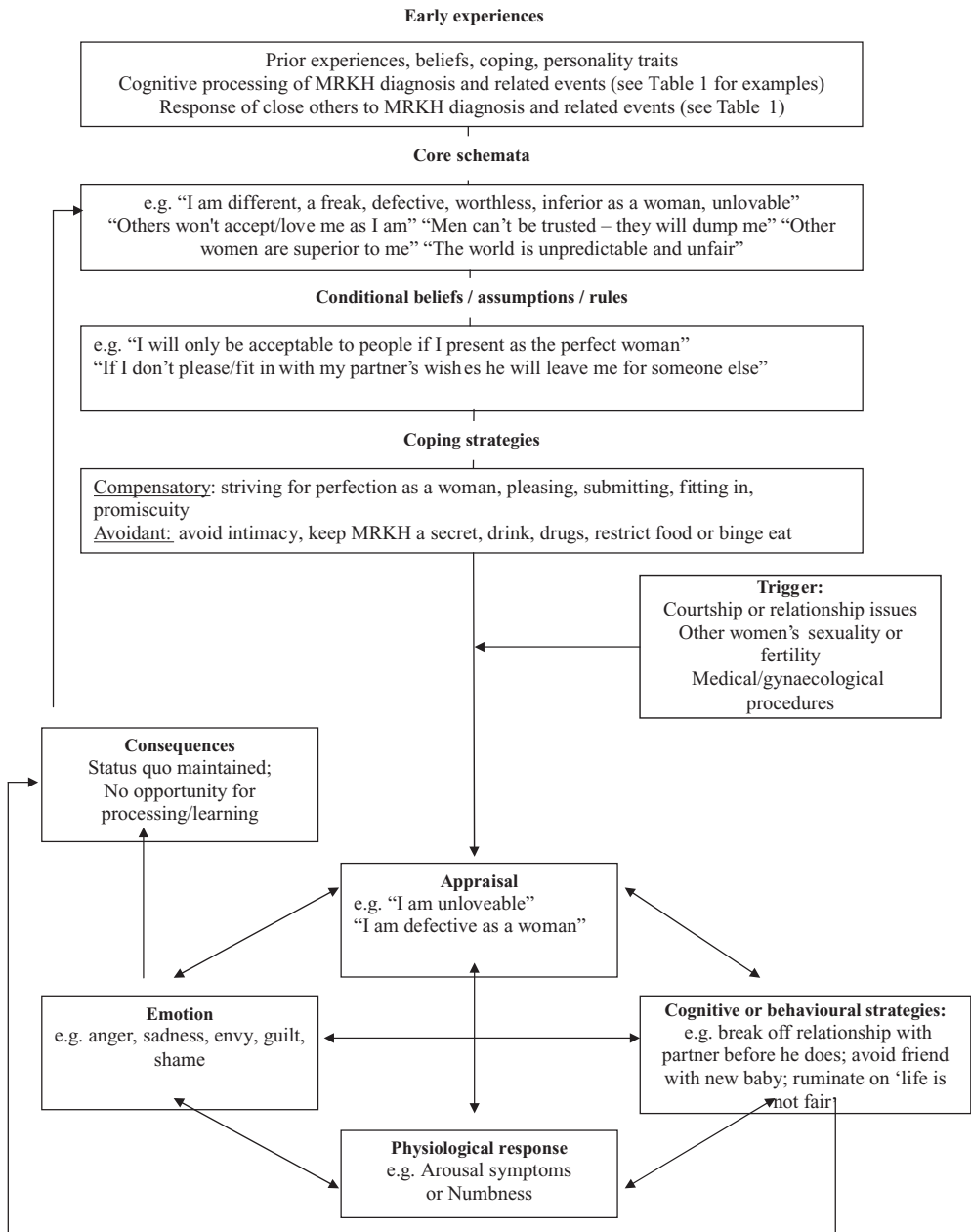


Figure 1. Conceptualization of problems in adjustment to MRKH

associated misconceptions to a more manageable and coherent evaluation of the experience. This work involves getting women to expose themselves to feared and avoided emotions and thoughts, identifying and challenging unhelpful appraisals, and systematically confronting and reappraising the event, challenging the negative conclusions drawn about themselves, others and the world, and core beliefs and rules or assumptions developed from it, or earlier beliefs about the self that are reinforced by it.

Cognitive restructuring and decentering. In later sessions other MRKH related events and problems (e.g. dilator treatment, issues around sexual and romantic relationships and infertility) are used to help women to process previously avoided thoughts and feelings, to identify and challenge unhelpful beliefs and behaviours and to develop a more adaptive view of themselves, the world and their future (Janoff-Bulman, 1992; Rothbaum and Fao, 1996). Moreover, there is also an attempt to help women find meaning in their experience. This involves acknowledging the real possibilities of both good and bad things happening, of MRKH being a source of strength not of weakness in oneself, of personal triumph and mastery, of relishing the good life, notwithstanding its shortcomings, and emerging from the experience somewhat sadder but wiser (Janoff-Bulman, 1992). It also involves letting go of the maladaptive schemata about the experience and regaining a sense of wholeness, and integrity, through a process of reassembly, where the confusion regarding the self is cleared and the interruption to body image, psychosexual differentiation and its varied parts is overcome and the body, the self and the condition cease to be an ongoing source of anxiety.

Dealing with maladaptive coping strategies and building new skills. Participants are also taught to identify unhelpful coping strategies that often make a person feel better in the short-term (e.g. starving or comfort-eating, promiscuity, pleasing, pretending that everything is “more than normal”, or using drink or drugs) but have negative longer-term consequences and replace them with more adaptive strategies. These include expressing difficult thoughts and feelings through sharing them with a close other, writing about them or using self-soothing strategies. Moreover, there is also a focus on building interpersonal skills, particularly in romantic and sexual relationships, e.g. how to mention MRKH to a new partner, when and what to tell, how to look after one’s interpersonal needs in a relationship, and how to have mutually satisfying sexual experiences.

Building new and more adaptive self-schemata. To help participants strengthen the alternative/more adaptive self schemata developed in therapy guided imagery is used: At the end of each session a guided imagery tape is played that follows the steps of: tuning into the body, creating a wise and compassionate figure, letting go of the negative images, reinforcing the alternative self schema, as described in Garner and Garfinkel (1997) and adapted to MRKH.

Furthermore, participants are asked to keep a positive data log of observed events or activities that they have engaged in that were in keeping with/reinforcing of their newly formed schema of themselves. The aim is to focus on the positive, away from the negative, and to slowly make this newly acquired “head-knowledge”, a “heart knowledge”, i.e. something that they really believe in about themselves without needing to question it.

Therapeutic writing tasks. In between sessions this work is supported by specifically designed homework tasks that use the tool of therapeutic writing. There is a large and sound body of research supporting the use of writing about traumatic or negative life events as a

tool for processing these (see Pennebaker 1997, 2004; Nicholson, 1995; Lange, 1994, 1996; Schoutrop et al., 1997). As MRKH is often a shameful secret for women that they do not find easy to talk about, it was thought that writing might be a useful way of helping them process and make sense of what had happened to them. A number of MRKH related writing exercises were constructed based on Pennebaker (2004). (Details can on request be obtained from the authors).

Evidence supporting the model and treatment

Broad support for this model and treatment comes first from a cross-sectional survey that compared women with MRKH ($N = 66$) with healthy control women recruited from the community ($N = 31$) of comparable age and socio-economic background (Heller-Boersma et al., in press) and second from the first ever randomized controlled trial of psychological treatment in women with MRKH conducted by our group and comparing the new CBT-MRKH group treatment with waiting list (Heller-Boersma et al., 2007).

On the Impact of Events Scale (Horowitz, Wilner and Alvarez, 1979), which measures symptoms of intrusion or avoidance after a major traumatic life event or life change, women with MRKH had mean scores in the moderately severe (clinical) range when asked to think about being diagnosed with MRKH as the event (Heller-Boersma et al., 2007). Women with MRKH were significantly more psychologically distressed than controls on the Symptom Checklist SCL-90-R (Derogatis, 1992; Heller-Boersma et al., in press). The scores of MRKH women were between population norms for non-patients and psychiatric outpatients (Derogatis, 1992). Together this lends some support to the notion that there was failure to “process” the diagnosis on a spectrum similar to what happens in PTSD.

Women with MRKH also had significantly lower self-esteem than control women on the Rosenberg Self-Esteem Scale (RSE; Rosenberg, 1965), although the mean RSE scores for both groups were in the normal range, and had significantly higher (pathological) scores on the Eating Disorders Inventory total score and several of the sub-scales (EDI; Garner, Olmstead and Polivy, 1983; Heller-Boersma et al., in press). The EDI measures both disordered eating behaviours and related cognitive and emotional aspects. Three of the subscales on which the MRKH women performed significantly poorer than control women related to self-efficacy and self-worth (i.e. “ineffectiveness subscale”), the ability to form close trusting relationships (i.e. “interpersonal distrust” subscale), and the inability to identify emotions and physical needs and sensations, such as hunger or satiety (i.e. “interoceptive awareness” subscale). In addition, one of the subscales on which MRKH women had significantly higher scores than controls was the “bulimia” subscale, which relates to overt eating disorder symptoms. These findings lend support to the notion that women with MRKH have a negative view of themselves, and may avoid intimacy and emotions and resort to maladaptive strategies, such as binge eating, to cope with their difficulties.

In the RCT (Heller-Boersma et al., 2007), 39 women with MRKH were randomly allocated to group CBT ($n = 19$) or waiting-list ($n = 20$). Outcomes were assessed at pre-treatment, post-treatment (7 weeks) and at 3-month follow-up. Women who received group CBT-MRKH, reported significantly reduced psychological distress, improved self-esteem, and a lessened impact of diagnosis at the end of treatment, with further gains at follow-up, whereas women on the waiting list remained unchanged. Effect sizes were medium or large.

Additional validation of the model comes from the therapeutic writings of RCT participants allocated to CBT-MRKH. Table 3 shows some typical excerpts from participants' homework writing assignments, illustrating the profound impact that MRKH has had on their lives at various stages and the change in their views as they begin to question unhelpful beliefs during therapy.

Table 4 shows some illustrative quotes from participants' "goodbye letters" describing their experience of therapy. These suggest that participants valued the group support, but also the specific techniques used, such as the therapeutic writing tasks and the cognitive restructuring of extreme and unhelpful beliefs. The quotes also illustrate the marked positive changes these women experienced in terms of their view of themselves and their MRKH.

Discussion

This paper describes a novel cognitive-behavioural conceptualization of how a problematic adjustment to MRKH arises in a particular developmental context and how this is maintained, and a cognitive behavioural treatment approach based on this model. The strengths of the model are that it is underpinned by a systematic review of the MRKH literature and validated by two empirical studies including both quantitative and qualitative data. The iterative process involved was based on the MRC framework for the development of complex interventions.

A novel feature of our model is that it takes into account the developmental context in which the events of MRKH typically unfold, in terms of these young women's cognitive and emotional development and the typical developmental tasks that they have to negotiate. This is something that might be of interest to researchers and clinicians working with conditions arising in adolescence.

However, further research is needed to refine the model and treatment. Our own studies, on which the model is based, are limited by the fact that they are based on women from an MRKH register in a specialist tertiary care centre, i.e. they include mainly women who have been diagnosed many years earlier. To the best of our knowledge there are no prospective longitudinal studies of psychological adjustment of women with MRKH. Such studies would give a more nuanced insight into the kind of psychosocial problems that develop and might also shed more light on who is particularly at risk of developing problems and what factors might reduce vulnerability and foster a more adaptive adjustment (Moorey, 1996).

Our RCT, which compared group CBT for MRKH with waiting list, found that women receiving the active treatment made lasting improvements. However, without adding a comparison intervention to this, consisting of an unstructured support group, we cannot be sure that it was the active ingredients of the intervention that made a difference or whether it was just non-specific factors such as re-moralization and group support that helped. Patients' own writings suggest that they found some of the specific techniques used helpful and something to fall back on at times of future difficulties.

MRKH is a relatively rare condition, and the question may therefore arise of what the significance is of this work for a wider audience. However, the model of treatment presented here is applicable or adaptable to a number of other congenital or acquired gynaecological conditions, such as premature ovarian failure, breast cancer, early onset endometrial cancer, female genital mutilation, Turner's Syndrome, ovarian dys/agenesis or Complete Androgen Insensitivity Syndrome, all of which have a psychological impact not dissimilar to MRKH in terms of women's sense of self and femininity. Thus, the model and

Table 3. RCT participants' homework writings about their experiences and therapy

Experience	Patients' writings
Run up to diagnosis	<p>a) My best friend had started her periods . . . I felt like I had been left behind, she was no longer the same as me she had become different. I waited but nothing happened to me . . . My period became the event everyone was waiting for but never happened. I had the feeling my mum and aunty were discussing it regularly . . .</p> <p>b) My younger sisters had started their periods. I was not worried, I thought periods happen only to some girls and not to others. . . . during the investigations I started worrying . . . that I had some disease and that I will die if I did not have any (periods) . . . no one explained . . .</p>
Being diagnosed	<p>a) She walked up to the side of the bed not sitting down to make me or her feel comfortable . . . I can't remember my parents being there, but they must as they heard the immortal words which she uttered from her lips "you can't have children, you can't have sex, you don't have a womb or vagina, if you have any questions ask the nurse". Turning on her heel she walked out with a very straight back. I don't remember much else after that about the day . . .</p> <p>b) . . . I was numb, and did not hear the rest of the conversation, after the explanation of MRKH, not having an uterus, I was just staring at him and did not ask any question, and started crying, the whole implication of MRKH became vivid in mind, at that point my world crashed, shattered, broke in pieces. I was inconsolable . . . I was devastated . . . I just wanted to die . . .</p>
Coping after diagnosis	<p>a) I blocked out what the doctor had said and hoped that my womb was only underdeveloped and would grow or that womb transplants would become available as a technique. I protected myself from reality, until I went again and it all started to sink in. I had wanted to hit the doctor, tell him that he was wrong, and had no right to determine my future. I became depressed, but then I decided to become the best I could and fluctuated from working very hard to being depressed and overeating and totally losing control, and wondering what it was all for . . . I avoided dad and the topic of (arranged) marriage.</p> <p>b) Amongst my friends and then colleagues at work I pretended to everyone that I was normal . . . During the day I was busy at work, going out in the evening. At night-time lots of questions would enter my head. Why me? Why was I born female, am I female if I can't do what other females do . . . How was I going to tell a future husband/partner . . . I decided subconsciously, I think now, that this was something I could never find answers to, I had to forget it – box it up, go into the world as a normal woman, and believe it wholeheartedly. Never tell anyone not even close friends . . .</p> <p>c) I finished with my boyfriend, I could not see a way of pursuing a relationship, as in my head I kept hearing the doctor's words (that it would take a special man to be in a relationship with me). I stopped going out, I stopped eating, I just worked. I exercised to get a perfect body and made myself as pretty as I could, to be accepted, as I felt so rejected . . .</p>
Conclusions drawn about self, others, the world, the future	<p>a) I felt everything had changed for me – I had always always wanted children – suddenly there was an empty space inside me which could never be filled. . . . I concluded I am different, strange, a freak, unfinished, incomplete, useless, ugly, shameful, unfeminine, unable to have something that is so important to me . . . unable to fulfill an important function – having children, . . . an outsider looking in on the rest of the "normal" world. . . . I am alone,</p>

Table 3. Continued.

Experience	Patients' writings
	<p>insignificant, . . . suddenly all grown up but too young and childish to deal with something of this magnitude . . . Others are lucky, normal, indifferent . . . unaware, ignorant, and different to me . . . They will never understand how I feel and I will never understand how they feel . . . The world is spiteful, cruel, stupid . . . empty, meaningless, it goes on in spite of everything.</p> <p>b) I am less important, an extra weight on this life and my family. Everything that has meaning has fallen to pieces, I am an in-complete woman, without any goals, a hopeless person, some-body that wastes space, an extra mitten. I am nothing I have nothing, no real value, nothing I do would count. The world has come to a dead end, there is no future, no way to go forward . . .</p> <p>c) Suddenly the goal-post had been moved. I wasn't what I thought I was. If felt apart from everyone, singular, unique. The world is a place where two identities meet and reproduce, I was not part of this world. But no one knew this but me. I could move within it unnoticed for what I was. I felt in disguise, someone masquerading as a girl/woman. But inside I felt different. I could not image the future it was a bleak hole . . .</p> <p>d) I was barren, a freak, someone who should not have been born, a miscarriage survived. Everyone would know that I was a freak . . . I was unloveable and inadequate . . .the plan of marrying and having babies was shattered.</p>
Challenging negative or idiosyncratic conclusions about the self, others, world and future	<p>a) I am still loveable, a bloke can take me or leave me, as I can offer him and the world much, beyond childbearing. I am looking for an advantage in this, working out a future with a different meaning and have much to achieve. This (MRKH) has not changed the character of the person I am, who is caring and responsible, a real fighter and peacemaker for the good of the world. I am being tested, and through this I have become more open minded, wiser, and stronger.</p> <p>b) I now perceive MRKH as any other syndrome that exists in the world, it is not a parasite anymore. In a way I am glad to have known, experienced and live with it, otherwise I would have also been ignorant . . . It made me a stronger person with a different view of life . . . I used to be a typical girl who would dream of getting married, having children . . . but now I have a different vision, I have MRKH, it does not disable me physically, I re-prioritize my plans, life, plan other things, enjoy everything more, than had I not had MRKH! It has enabled me to deal differently with others, I used to be a very passive and wanting to please everybody type of person. I realized that I am harming myself . . . and decided to stand up for myself, I, literally, do not worry about others' opinion of me anymore, because I think highly of myself which I never used to do . . . It does not mean that MRKH has not been the biggest loss in my life in terms of grief . . .but it has also opened things up for me . . . I see things I otherwise would not have seen . . . I can be myself, improving and becoming the character I used to be but with also the added one fortified by MRKH! I can still become what I want to become with MRKH alongside me!</p> <p>c) I ask myself what is the worst that can happen and MRKH is not that bad in the scheme of things. It effects small parts of my life, which I could address if I so wish. I am not sick, I am not disabled, I am not stupid, or ugly, I have eyes and I can see and understand . . . and look deeper . . . and if others do not that is their problem. I am strong, I can cope with anything. I focus on what is important in my life . . . look after myself and live life day by day. I do not need to prove myself and relationships I can take or leave.</p>

Table 4. Participants views of different aspects of the treatment and on how treatment has altered their view of themselves

Patients views of different aspects of treatment	
Social support	<p>(a) The first time that I could talk and share this terrible secret. I could do it in a safe way . . . I was allowed to be upset, and no one was shocked by what I had to bring, I could be honest, and I was believed.</p> <p>(b) “I feel liberated by discovering that others have had similar experiences and reactions to me . . . Now I can judge myself less harshly and feel less inclined to hide myself from other people, as I discovered that I am more sane and normal than I thought. Sharing our stories the weight of loneliness has shifted from my shoulders and mind, and I feel more positive, and supported”.</p> <p>(c) “I will miss coming, but I now associate MRKH with this room of people with whom I have forged links and have shared so intimate detail, . . . To know that you are all out there brings me so much comfort; . . . I will always have you all in my head and draw strength and knowledge from you all, knowing that you are there maybe feeling similar things.”</p>
Writing tasks	<p>(a) “Through the writing exercises, and discussing these in therapy, I have been provided with a powerful tool that has allowed me to reorganize the somewhat muddled memory of being diagnosed with MRKH. . . .a huge problem has been broken down, making it more manageable, and a smaller “monster” which plays a smaller role in my life than it previously did . . .</p> <p>(b) What I truly enjoyed and found helpful have been the writing exercises. I express my emotions better in writing than verbally, and by doing so, it has helped me to reflect back on where I have been to, where I am now, looking within myself and pulling out more strengths and qualities that I did not think existed, learning that asserting myself is a good thing. Also by writing those exercises, it is something that I can look back or read when maybe I am feeling low or just need encouragement, a reminder of what makes me a woman!!!</p>
CBT strategies – challenging unhelpful beliefs	<p>(a) “By getting varied perspectives, opinions have been turned on their head, making you consider other issues around it, and discover different and new dimensions. Through the process of coming to understand ones’ thoughts and feelings and challenging these, the negative image of MRKH and its varied aspects have been turned round.”</p> <p>(b) I challenged the messages that I had picked up from my family and the doctor and which did not necessarily get shaped into the right beliefs, as it was not as simple as “loved, not loved”. . . .Therapy has made me feel like this; I am loveable as a person I am good enough, you don’t need a womb or children for that</p> <p>Impact of therapy on patients’ views of themselves and their MRKH</p>
Increased acceptance of MRKH	<p>(a) “MRKH does not need to be the be all and end all of my experiences, or the heavy load that will stand in the way of everything. It can be put in “its place”, it need not dominate my life. . . .I will live life to the full, no matter what happens. I can now bring back color and choices in my life, reach out for my dreams and make a difference to myself and others. I . . . am more accepting of MRKH . . . , both in my heart and mind.”</p>

Table 4. Continued.

Patients views of different aspects of treatment	
	(b) "I have allowed myself to acknowledge MRKH rather than bury it. Life has made me naturally brave, but now I am being braver by acknowledging that I can have feelings about MRKH and express these. I am learning to "mourn" the choices that I would have faced in a different way had I not had MRKH, mark these, and make choices from those available to me and my husband."
Shedding MRKH as identity	"I feel a great sense of relief, more free, facing a future that feels less burdened, brighter; with much shame, embarrassment and an "ugly" secret "dispersed". I now know that MRKH is not me, I am not MRKH; it is just a part of me, but I am also much more than that. . . . I can now see that having the condition does not make me anything of a lesser person, I am as lovable, acceptable, and capable as the next person, and people will like me for who I am . . . I can interact with people differently and say "no" when I need to, and be honest about myself to others, I no longer have anything to hide."
Reducing the power of MRKH	"To confront this condition has been to break its power over me. As I come to terms with the enormous significance of this realization, it is like music to my soul. I am free to break out from the bars behind which I have been trapped for so long, now I can fly . . ."

therapy described are of relevance to CBT practitioners working in a broad range of clinical settings from general practice through to different medical/hospital settings (paediatrics, gynaecology, endocrinology) and in other clinical psychology, psychiatry or mental health settings.

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