Practice case study

KEYWORDS MAYER-ROKITANSKY-KÜSTER-HAUSER SYNDROME | REFERRAL | SPECIALIST CENTRES

Nurses are constantly dealing with new and challenging situations. Case studies are a way of sharing these experiences and offering possible solutions.

Exploring the support options for patients with Mayer-Rokitansky-Küster-Hauser syndrome

BACKGROUND

Girls with Mayer-Rokitansky-Küster-Hauser syndrome (MRKH) are born without a vagina or uterus. It affects one in 5,000 female births (Aittomaki et al., 2001). They have 46XX chromosomes, normal ovaries and secondary sexual characteristics but present with primary amenorrhea because of uterine absence.

The aetiology is unknown. There are two forms: uterine and vaginal absence; and, another type associated with other congenital malformations.

PATIENT HISTORY

Sally-Ann* had not started her periods by the age of 15, although her secondary sexual characteristics developed normally. Women in her family had their menarche at 12–13 years. Her GP said that Sally-Ann was a ‘late starter’ and prescribed hormonal treatment.

A year later, nothing had happened so the GP referred her to their local gynaecologist. A laparoscopy found uterine and vaginal absence but the presence of ovaries.

Referral to the specialist centre

The gynaecologist referred Sally-Ann to our national centre for reconstructive surgery. The specialist here explained MRKH in detail to Sally-Ann and her mother. He also informed them that uterine transplants are still experimental, which they found devastating. But it is vital to be honest, no matter how painful the truth, as patient trust in clinicians is crucial. Vaginal dilator therapy is the first-line treatment for absent vaginas (American College of Obstetricians and Gynaecologists, 2002).

I saw them after their consultation. At the centre we ensure that patients understand fully what they have been told. They can ask questions or express their feelings, and I can further explain their treatment.

At this visit, Sally-Ann appeared depressed and her mother was also upset. It was important to help them put things into perspective to ensure they had a better understanding. I explained that Sally-Ann is female and reiterated what was normal and abnormal, and what we could and could not do. She explained that she felt embarrassed, confused and lonely. I told them that we have about 500 girls with MRKH at our centre.

Sally-Ann was told about support available – an MRKH support group, one-to-one patient contacts, a helpline and online support through a website and chat room. Fertility, IVF surrogacy and adoption were also discussed.

Preparing for treatment

I explained dilator treatment, showed Sally-Ann some dilators and encouraged questions. As I was aware of anxiety and the possibility of information overload, I provided information booklets on MRKH, dilator therapy, adoption and IVF surrogacy.

Sally-Ann saw our psychologist to help her cope and come to terms with her condition. It is best to start treatment when patients are ready, to ensure adherence. Most start when they are in relationships, although some prefer to start before so they feel confident.

Treatment

When Sally-Ann turned 18, she was admitted into hospital for dilator therapy (10–15 minutes for each treatment, three times daily). The correct application of firm pressure and progression of dilator sizes are necessary to create an anatomically functional vagina. Patients need encouragement and reassurance, so part of my role is the one-to-one support, teaching and supervision of dilator treatments.

After three days, Sally-Ann was confident and discharged to continue her treatment at home. She was reviewed two weeks later and thereafter 4–6 weekly until her vagina was normal at 7–8cm (Lloyd et al., 2005).

Reviewing patients regularly until treatment is completed is important, as this motivates and ensures adherence and gives them the opportunity to discuss any issues. Sally-Ann completed her treatment within three months.

CONCLUSION

Sally-Ann attended our support group meeting and found it beneficial talking to others. She is in a stable relationship and feels normal.

These patients should be referred/treated at centres with the expertise and resources that embrace a multidisciplinary team approach.

* The patient’s name has been changed.

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REFERENCES


Have you been involved in a new treatment or therapy?
Have you been involved in a situation that has made you think about or change your practice?
If you or your colleagues would like to share your experience email your ideas to mti@emap.com, putting ‘case studies’ in the subject box.