Received: 29 January 2024 | Revised: 15 March 2024 | Accepted: 24 March 2024

DOI: 10.1002/ccr3.8732

CASE REPORT

Clinical Case Reports Open Access WILEY

Bilateral indirect ovarian inguinal hernia in a young female with type 1 Mayer–Rokitansky–Küster–Hauser syndrome: An extremely rare clinical context

Sunil Bhatta [1] | **Hemant Ojha** [1] | **Meraj Alam Ansari** [2] | **Anuj Shrestha** [3]

[1]Department of Anesthesia and Critical Care, Nisarga Hospital and Research Center Pvt. Ltd, Dhangadhi, Nepal

[2]Department of General Surgery, Nisarga Hospital and Research Center Pvt. Ltd, Dhangadhi, Nepal

[3]Department of Radiodiagnosis and Imaging, Nisarga Hospital and Research Center Pvt. Ltd, Dhangadhi, Nepal

Correspondence
Sunil Bhatta, Department of Anesthesia and Critical Care, Nisarga Hospital and Research Center Pvt. Ltd, Dhangadhi, Nepal.
Email: bhattasunil26@gmail.com

Key Clinical Message

Incidence of bilateral inguinal hernia encompassing bilateral ovaries in adult female is very thin and concomitant association with Mayer–Rokitansky–Küster–Hauser syndrome is out of ordinary. Along with surgical management of hernia, these females need multidisciplinary slant to manage gynecological, social, and emotional issues.

Abstract

In mature females, bilateral ovarian inguinal hernias are a rarity. In this situation, ultrasonography is the basic adjunct to confirm the diagnosis. Mayer–Rokitansky–Küster–Hauser syndrome is typically linked to ovarian hernias in grown-up females. The most important ways to avoid problems are early diagnosis and surgical repair. A 25-year-old lady presented to our outpatient clinic with a history of swelling in bilateral inguinal region for 1 month. On the ultrasound examination, the right ovary was visualized in the right high inguinal canal, and the left ovary was seen at the level of deep inguinal ring with no visualization of the uterus in its normal anatomical position. The patient underwent bilateral inguinal exploration under spinal anesthesia, and herniated contents were successfully reduced back to anatomical locations. Clinical care for such a clinical condition must be multifaceted, involving intensive counseling, relocating the uterus, fallopian tube, and ovary to preserve fertility, and preventing consequences like incarceration and strangulation.

KEYWORDS

hernia, karyotype, Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome, ovaries

1 | INTRODUCTION

Inguinal hernia is exclusively common in males and relatively uncommon in females. Mostly, the small intestine and omentum protrude into the hernial sac. In extremely rare instances, adult females develop inguinal hernias encompassing the uterus, fallopian tubes, and ovaries. As many as 31% of inguinal hernia sacs in young girls involve uterine adnexa; bilateral ovarian sliding hernias are unusual, and their incidence declines with age, making it infrequent in adult women. A substantial number of reported cases in adult females have been linked to additional genital tract congenital abnormalities.[1–4]

Mayer–Rokitansky–Küster–Hauser (MRKH) syndrome is a moniker assigned to the occurrence of Mullerian dysgenesis in the ovaries, uterus, and fallopian tube. The absence of paramesonephric ducts leads to halt in normal development of uterus, cervix, and vagina. However, the ovaries and fallopian tube have normal development; karyotyping reveals normal 46, XX and females exhibit signs of normal thelarche and pubarche.[2,5]

There are two reported subtypes of this syndrome: Patients with type 1 have malformations pertaining to the genital system; meanwhile, patients with type 2 have an aberrant MURCS (Mullerian, Renal, Cervicothoracic Somite) association abnormality.[2,3]

In this article, we document the case of a 25-year-old female with type 1 MRKH syndrome. She initially presented to our outpatient department with a complaint of swelling in the bilateral inguinal region.

The majority of clinicians are not familiar with MRKH syndrome because it is seldom seen in clinical practice. Introducing this infrequent case is our primary objective.

2 | CASE HISTORY AND EXAMINATION

A 25-year-old lady presented to our outpatient clinic with a history of swelling in the bilateral inguinal region for 1 month. Her prior health records were normal, and she had no history of surgery or trauma. Most importantly, she stressed not having had a single episode of menstrual cycle up to that age. There were no recognized medical conditions in her family history.

She was 1.60 m tall, and coarse hair was present across pubis, medial thigh, and arm pits. Adult breast contour was noted with flat areola. On abdominal examination, palpable swelling were noted in the bilateral inguinal region. Both of the swellings were reducible and demonstrated a positive cough impulse test. The examination of the cardiovascular, respiratory, and nervous systems was unremarkable.

2.1 | Investigation

Based on examination findings, an ultrasound scan of the abdomen and pelvis was requested. On the ultrasound examination, the right ovary was visualized in the right high inguinal canal (Figure 1), and the left ovary was seen at

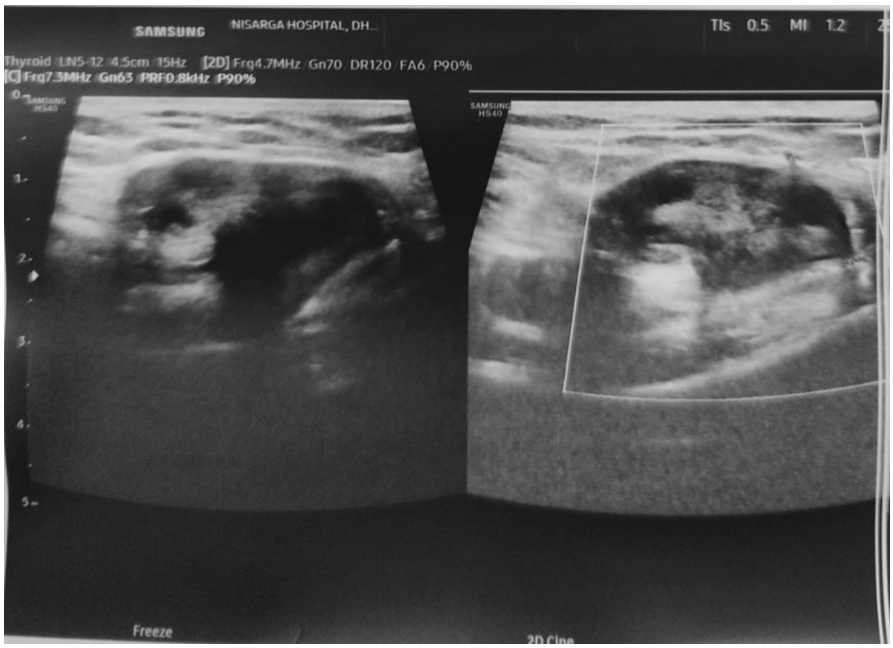

FIGURE 1 Right ovary with normal vascularity in right the high inguinal canal.

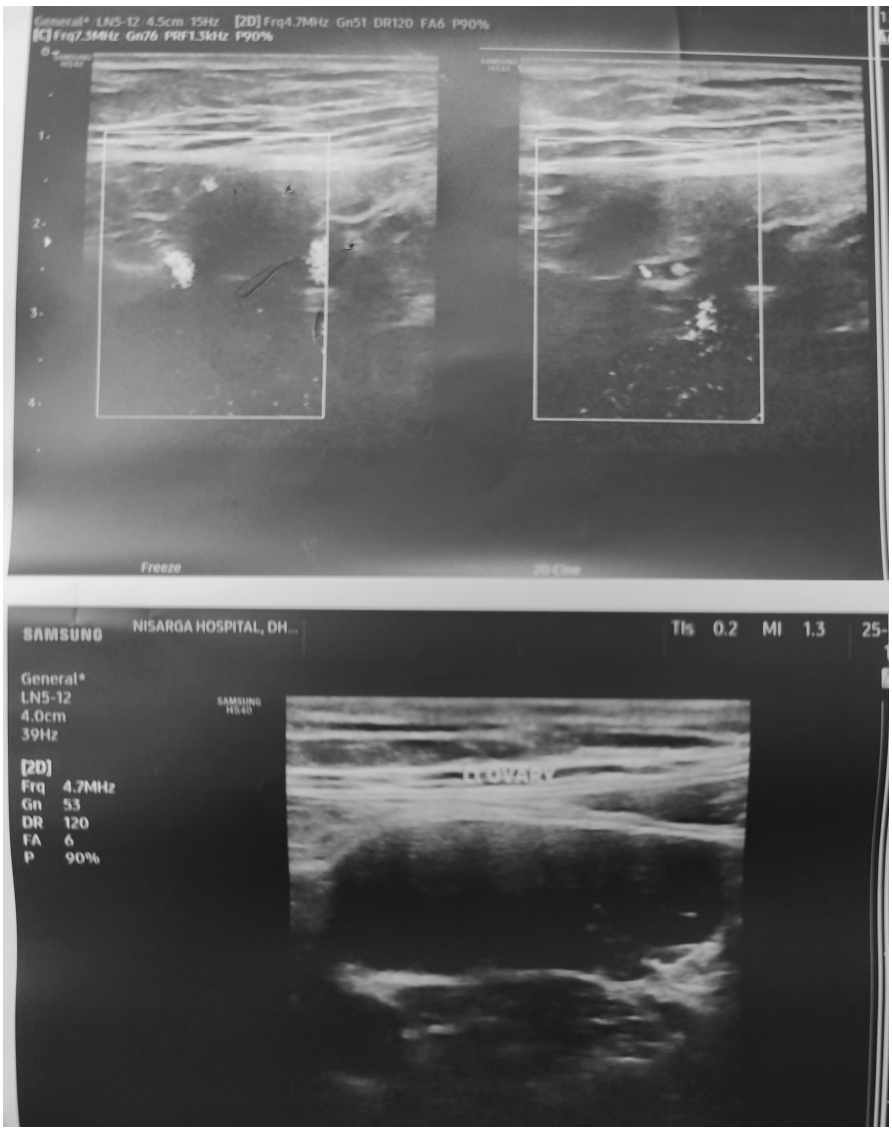

FIGURE 2 Left ovary at level of the left deep inguinal ring with normal vascularity.

the level of the deep inguinal ring (Figure 2). Both of the ovaries were normal in size. Additionally, the uterus was not visualized in the scan, suggesting uterine agenesis or severe hypoplasia (Figure 3). No abnormalities in other abdominal organs (liver, kidneys, bladder, and spleen) were noted. Based on the ultrasonographic findings, a sample of blood was sent for karyotyping, which later came out to be 46, XX.

2.2 | Diagnosis

A clinical diagnosis of type 1 MRKH syndrome with bilateral ovarian inguinal hernia was made. Hence, the patient and her guardian were educated in detail about the nature of the condition and the need for inguinal ovarian hernia repair to prevent irreducibility and incarceration that could possibly occur.

2.3 | Management

With proper written and informed consent, the patient underwent surgery under spinal anesthesia. An indirect inguinal hernia was discovered during intraoperative dissection of the right inguinal canal. The hernial sac was opened anteriorly, unveiling the ovary, right fallopian tube, and rudimentary uterus (Figure 4). The hernial contents were reduced with extreme caution, and a herniotomy was performed. In the end, mesh plasty was employed to reinforce the posterior wall of the inguinal canal (Figure 5). The same surgical technique was deployed to dissect the left inguinal canal. Intraoperatively, the hernial sac was found to contain the left ovary and fallopian tube (Figure 6). Hernial contents were reduced, and herniotomy along with mesh hernioplasty was performed (Figure 7). The patient had an uneventful perioperative course and was discharged on postoperative Day 3.

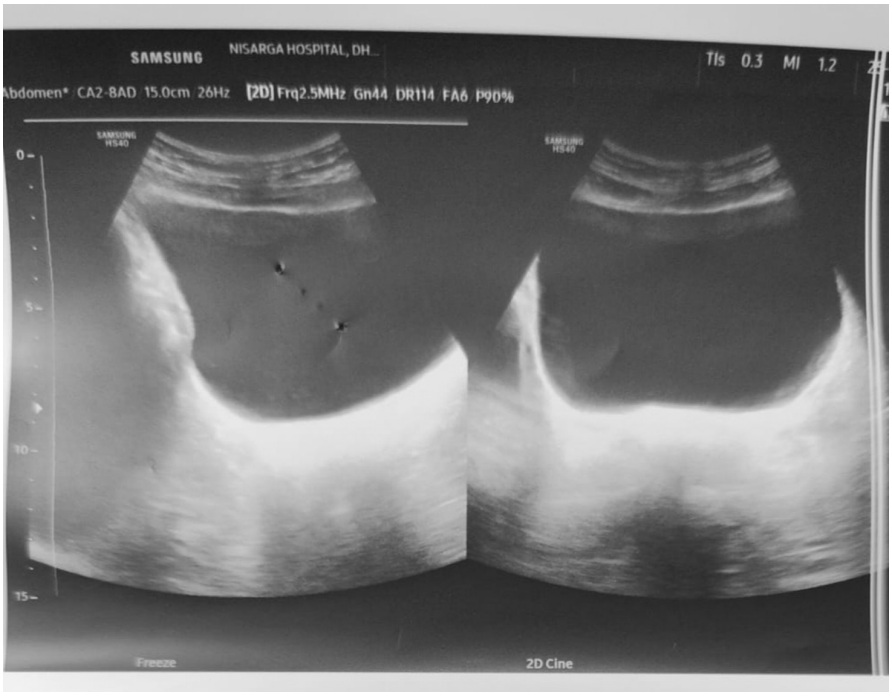

FIGURE 3 Ultrasound of the pelvis with no visualization of the uterus in its normal anatomical position with a full urinary bladder.

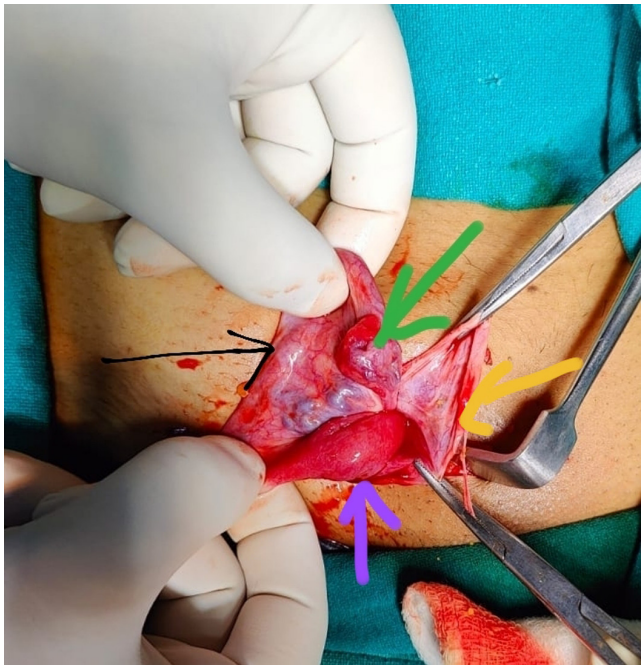

FIGURE 4 Picture depicting the right hernial sac (yellow arrow), rudimentary uterus (purple arrow), right fallopian tube (black arrow), and right ovary (green arrow).

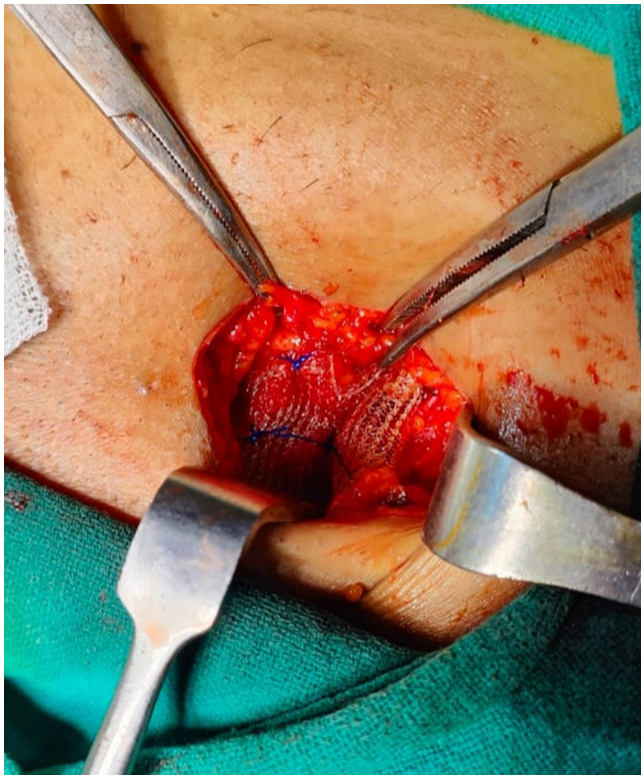

FIGURE 5 Mesh repair of the right inguinal canal.

Before discharge, the patient was also counseled regarding vaginoplasties and future fertility prospects from the perspective of the native administrative system, sociocultural system, and religious beliefs, and advised for regular follow-up in gynecology clinics for genital issues.

3 | DISCUSSION

Although inguinal hernia is less frequently seen in young female infants as compared to males, the most often stumbled upon structures in female infants inguinal hernia sacs are the ovaries, with or without fallopian tubes.[6]

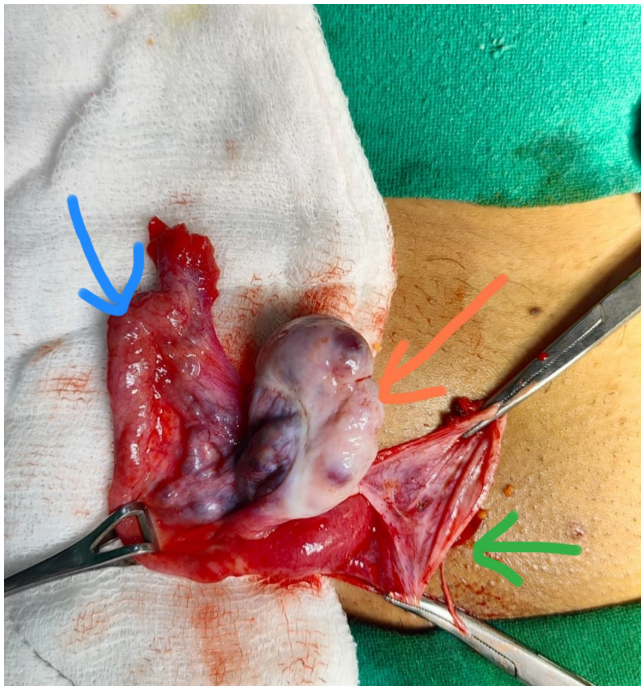

FIGURE 6 Picture showing the left hernial sac (green arrow), left ovary (orange arrow), and left fallopian tube (blue arrow).

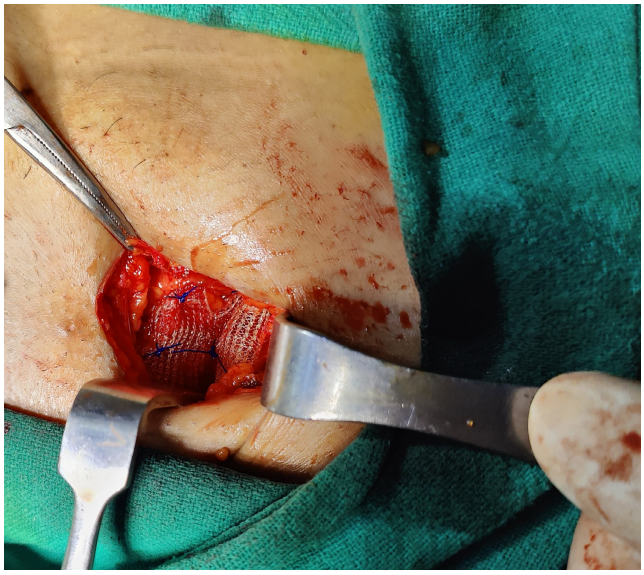

FIGURE 7 Mesh repair of the left inguinal canal.

However, it is exceedingly uncommon for ovaries to be found in the inguinal areas during adolescence and maturity. In these situations, Muller anomalies, which result from Muller aplasia and MRKH syndrome, are part of the etiopathogenesis.[6,7]

Initially situated inside the abdominal cavity, the reproductive glands subsequently descend. By the end of the eighth month of pregnancy, testes have become localized in the scrotum, and the processus vaginalis obliterates following birth. The nonobliterated processus vaginalis in females is known as the canal of Nuck and is a contributing factor to ovarian herniation, sometimes in conjunction with the uterine tubes.[2,8]

The incidence of MRKH syndrome is 1 in 4000–5000 live births and has an autosomal dominant mode of inheritance with incomplete penetrance and variable expressivity.[9] The aberrant development of the paramesonephric ducts is the primary cause of MRKH syndrome. Since the ovaries emanate from genital ridges, these patients have normal gonadal function. Upon impact on genitalia, the condition is defined as type 1 MRKH syndrome, whereas, type 2 MRKH or MURCS association syndrome includes renal anomalies (unilateral agenesis, ectopic kidney, horseshoe kidney), skeletal anomalies (fused cervical vertebrae, scoliosis, Klippel–Feil syndrome), and hearing loss or cardiac defects.[2,3,5,8,9]

Individuals with MRKH syndrome have a normal female habitus, including a typical thelarche and adrenarche. Primary amenorrhea is the very first symptom of MRKH syndrome in young women who otherwise present with normal development of secondary sexual traits, karyotype 46, XX, and no discernible chromosomal aberration.[5,9] In our case, the patient presented with swelling in bilateral groin of recent onset with a background history of primary amenorrhea.

It is obscure how genital organ herniation occurs in cases of groin hernia. According to Ozkan et al.[10] herniation may be spurred on by weakening of the ovarian suspensory ligament and broad ligament, which may worsen under conditions of increased abdominal pressure. Various theories are formulated by gallant researchers. Thomson's[11] theory states that there is a greater likelihood of the entire uterus, ovary, and fallopian tube herniating into the inguinal canal when the Mullerian ducts fail to fuse, which causes the ovaries to move overly. Conversely, Fowler[12] postulated that the fundamental cause of ovarian inguinal hernia was lengthened ovarian suspensory ligaments. Some also contend that the inguinal canal herniation of the ovaries was caused by ovarian or broad ligament weakening and an elevation in intra-abdominal pressure.[13]

Kumar et al. reported a case of bilateral ovarian inguinal hernia with left sided irreducibility in a 21-year-old female with Mullerian agenesis. Similarly, Samantroy et al. reported twisted gangrenous ovary with fallopian tube and underdeveloped uterus as the content of indirect inguinal hernia in a 34-year-old female with Mullerian agenesis in 2021.[1]

The patient had a successful emergency hernia exploration with a left oophorectomy, fallopian tube uterus relocation, and herniorrhaphy. Moreover, irrespective of congenital genitourinary conditions, multiple publications

have recorded inguinal herniation of the ovary with or without the fallopian tube and uterus.[14] Such as Okada et al. description of a case of a 1-year-old female girl with an irreducible inguinal hernia that covered the whole uterus, fallopian tube, and entire ovaries and Fowler's explanation of sliding inguinal hernia in a 1-year-old infant comprising bilateral ovaries.[12,13]

There are exceptionally few published circumstances involving ovarian herniation in MRKH syndrome, either with or without the uterus and fallopian tube. In 1980, Riggall and Cantor reported the first illustration of an inguinal hernia containing a uterus in a woman with a female karyotype, phenotypic traits, and infertility.[1,5]

Mohanty et al. and Bazi et al. reported a case of bilateral inguinal hernia containing ovaries in a 20- and 12-year-old female with features of MRKH syndrome.[5,15] Likewise, Saini et al.[3] also published a case report of 20-year-old lady with Mullerian defect who developed a right indirect inguinal hernia containing the right ovary. In addition, case reports of MRKH syndrome with inguinal hernia having ovaries as content are described in literature by various authors, namely Shumarova et al., Khan et al., and Yao et al.[1] While delving more into a piece of published work, a young woman with type 2 MRKH syndrome and a right utero-ovarian inguinal herniation who lacked a left kidney came across, which was described by Al Omary et al.[2] Type 2 MRKH syndrome has also been described by Jafari et al.[4] in a 12-year-old girl who developed inguinal herniation of the left ovary, fallopian tube, and left horn of underdeveloped bicornuate uterus along with ectopic kidney.

Ovarian torsion, blockage, irreducibility, strangulation, and gangrene are the common repercussions of ovarian inguinal hernia. About 4%–37% of complex cases of inguinal hernia have an irreducible ovary, whereas 2%–33% of cases have torsion or infarction.[1] The most important steps to prevent complications and restore fertility are early diagnosis and treatment. Independent of Mullerian status, relocation and herniorrhaphy are recommended as soon as the ovarian tissue in the hernial sac is identified.[1,2,6,7] Reduction and repair were carried out employing the open method on our patient. This repositioning can also be done laparoscopically.

Patients with MRKH syndrome require sophisticated care that involves a multidisciplinary approach centered on the patient, careful discussion, and treatment of all gynecological, sexual, mental, emotional, and reproductive difficulties. Through in vitro fertilization and surrogacy, it is possible for couples to establish genetic offspring. Two alternatives exist for performing sexual intercourse: surgically creating a neovagina or manually dilating the vagina with dilators.[2–5,14]

4 | CONCLUSION

In conclusion, we report an exceptional situation of bilateral simple indirect inguinal ovarian hernia in a female patient with type 1 MRKH syndrome. Infertility may ensue after torsion and subsequent infarction, rendering the ovaries doomed. When dealing with such clinical presentation, comprehensive care is required, which involves relocating the uterus, fallopian tube, and ovary, fixing the hernia, and opening up the neovagina. In particular, for young adult females, fertility preservation requires appropriate surgical and medical therapy.[1–15]

AUTHOR CONTRIBUTIONS

Sunil Bhatta: Conceptualization; formal analysis; resources; writing – original draft; writing – review and editing. **Hemant Ojha:** Writing – review and editing. **Meraj Alam Ansari:** Resources; writing – review and editing. **Anuj Shrestha:** Resources; writing – review and editing.

ACKNOWLEDGMENTS

None.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

No conflict of interests.

DATA AVAILABILITY STATEMENT

Data described to support the findings is openly accessible in the article.

ETHICS STATEMENT

This is a retrospective case report, and no sampling was used. The ethical approval can be waived.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID

Sunil Bhatta https://orcid.org/0009-0004-7871-3489

REFERENCES

1. Kumar A, Kumar A, Anwer M, Kumar D. Case of Mullerian agenesis presenting as bilateral inguinal hernia with left sided irreducibility in a 21 years old female: a rare case report. *Int J Surg Case Rep*. 2023;103:107895.
2. Al Omari W, Hashimi H, Al Bassam MK. Inguinal uterus, fallopian tube, and ovary associated with adult

Mayer–Rokitansky–Küster–Hauser syndrome. *Fertil Steril*. 2011;95(3):1119.e1-1119.e4.
3. Saini R, Bains L, Kaur T, et al. Ovarian inguinal hernia—a possibility in MURCS syndrome. *J Ovarian Res*. 2021;14(1):114.
4. Jafari R, Javanbakht M, Dehghanpoor F. Inguinal herniation of left ovary, fallopian tube, and rudimentary left horn of bicornuate uterus associated with type 2 Mayer–Rakitansky–Kuster–Hauser (MRKH) syndrome in a teenage girl: a case report and literature review. *Eur J Radiol Open*. 2020;7:100215.
5. Bazi T, Berjawi G, Seoud M. Inguinal ovaries associated with Müllerian agenesis: case report and review. *Fertil Steril*. 2006;85(5):1510.e5-1510.e8.
6. Khan WF, Rathore YS, Pol MM, Singh G. Inguinal hernia with ovary as content-laparoscopic repair in a Mayer–Rokitansky–Küster–Hauser syndrome patient: case report. *Int Surg J*. 2019;6(4):1421-1423.
7. Masoumi Shahrbabak M, Ebrahimi MF. A rare case of Mayer–Rokitansky–Küster–Hauser syndrome with right ovarian torsion and hypoplasia of the left adnexa. *J Surg Case Rep*. 2021;2021(5):rjab130.
8. Shumarova S, Maslyankov S, Gribnev P, Sokolov M. Inguinal hernia containing a fallopian tube and ovary with giant cyst in patient with Mayer–Rokitansky–Küster–Hauser syndrome. *Ann Clin Case Rep*. 2021;6:1995.
9. Prodromidou A, Machairas N, Garoufalia Z, et al. Ovarian inguinal hernia. *Ann R Coll Surg Engl*. 2020;102(2):75-83.
10. Ozkan OV, Semerci E, Aslan E, Ozkan S, Dolapcioglu K, Besirov E. A right sliding indirect inguinal hernia containing paraovarian cyst, fallopian tube, and ovary: a case report. *Arch Gynecol Obstet*. 2009;279(6):897-899.
11. Thomson GR. Complete congenital absence of the vagina associated with bilateral hernia of uterus, tubes, and ovaries. *Br J Surg*. 1948;36(141):99-100.
12. Fowler CL. Sliding indirect hernia containing both ovaries. *J Pediatr Surg*. 2005;40(9):13-14.
13. Okada T, Sasaki S, Honda S, Miyagi H, Minato M, Todo S. Irreducible indirect inguinal hernia containing uterus, ovaries, and fallopian tubes. *Hernia*. 2011;16(4):471-473.
14. Samantroy S, Mishra A, Panda J, Jena P. Ovarian inguinal hernia in premenopausal women: a case report. *Cureus*. 2021;13(12):e20846.
15. Mohanty HS, Shirodkar K, Patil AR, Rojed N, Mallarajapatna G, Nandikoor S. A rare case of adult ovarian hernia in MRKH syndrome. *BJR Case Rep*. 2017;3(3):20160080.

How to cite this article: Bhatta S, Ojha H, Ansari MA, Shrestha A. Bilateral indirect ovarian inguinal hernia in a young female with type 1 Mayer–Rokitansky–Küster–Hauser syndrome: An extremely rare clinical context. *Clin Case Rep*. 2024;12:e8732. doi:10.1002/ccr3.8732