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Case Report Open Access

Sigmoid Vaginoplasty in Patient with Complete Androgen Insensitivity: Technique and Outcomes

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Introduction

Androgen Insensitivity Syndrome (AIS), also known as testicular feminization, is a rare medical condition that has garnered increased attention in recent years due to evolving research and therapeutic approaches [1,2], as well as the psychological repercussions it has on patients and the risk of being associated not only with sex cord–stromal tumors but also with rare mesenchymal tumors [3]. AIS results from anomalies in the X chromosome and leads individuals with an XY genotype to exhibit various degrees of female physical traits or complete feminine characteristics. A genetically male individual who lacks sensitivity to androgens, resulting in the inability to respond appropriately to male sex hormones, is diagnosed with Androgen Insensitivity Syndrome (AIS). The condition manifests as an individual having one X chromosome and one Y chromosome but displaying female genital characteristics.AIS, is a condition pertaining to sex differentiation that affects the sexual development of male fetuses within the uterus and during puberty. It hinders proper development of male genitalia resulting in almost inevitable infertility upon adulthood. In this discussion, we will explore two distinctive cases of AIS and discuss the therapeutic approaches employed to manage this unique disorder, referencing recent developments in the field.

Abbreviations: AIS: Androgen Insensitivity Syndrome; AFP: Alpha Fetoprotein; CT: computed tomography; DSD Disorder of Sex Development HCG: human chorionic gonadotropin ; LDH: Lactate dehydrogenase; MRKH: Mayer-Rokitansky-Küster-Hauser

Keywords: Complete androgen insensitivity; Sigmoid vaginoplasty

Case Report 1:

Our first case involves a 23-year-old female patient, the third daughter in a family of four siblings, born from a consanguineous marriage. She sought medical attention due to primary amenorrhea, and notably, her two older sisters also experienced the same symptom.

Clinical examination unveiled a distinct female phenotype, characterized by the presence of axillary and pubic hair. Breast development was within the normal range, graded at Tanner stage 4. However, there was a conspicuous absence of a vaginal hiatus, accompanied by clitomegaly, a clearly visible urethral meatus, and labia majora hypertrophy. (Figure 1)

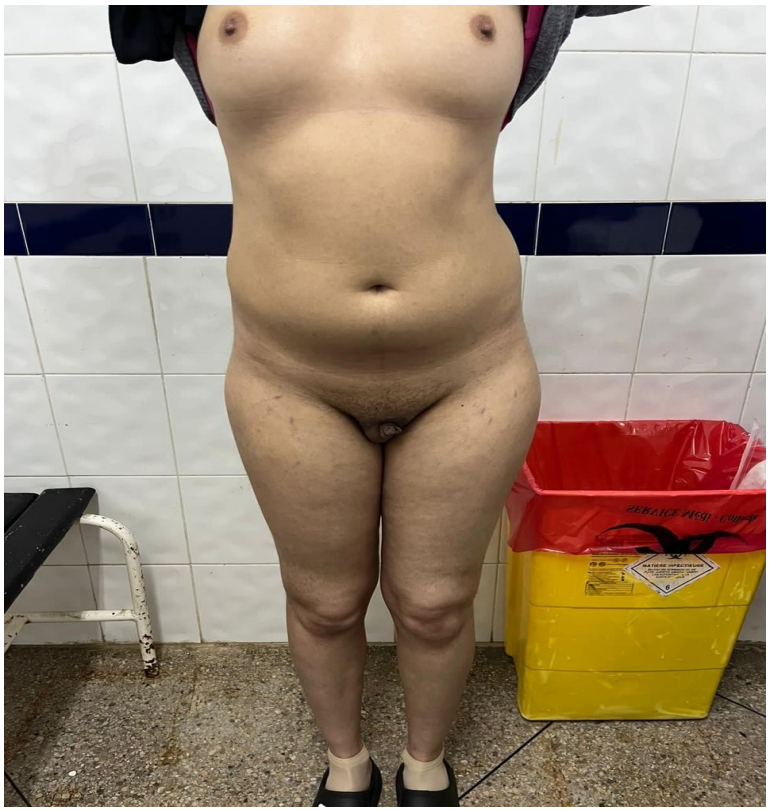

Figure 1: Female phenotype, with axillary and pubic hair, breast development was normal on Tanner scale 4

Palpation revealed two tender masses, one on the right at the level of the labia majora and another on the left in the inguinal region. Abdominopelvic ultrasonography detected two well-limited, oval, echogenic formations that were sensitive to compression. Notably, no vagina, uterus, adnexa, or prostate were identified. Computed Tomography (CT) imaging corroborated the presence of two nodular formations over the right vulvar region and the left inguinal canal. (Figure 2)

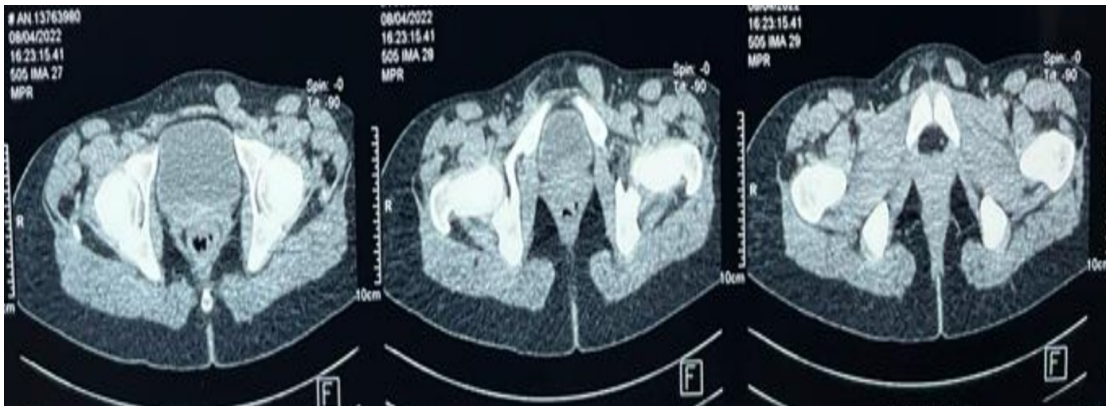

Figure 2: Sagital CT section showing no vagina, uterus, adnexa, or prostate were identified

Biological tests unveiled a testosterone level of 1.02 ng/ml, below the normal range for males, and FSH and LH levels of 2.06 mIU/ml and 18 mIU/ml, respectively. Additionally, estradiol (E2) levels were measured at 72.5 pmol/l, with tumor markers (AFP, HCG, LDH) returning within normal ranges.

Remarkably, an XY karyotype was identified, a stark contrast to the female morphotype observed. Intriguingly, sisters with a similar phenotype exhibited the same karyotype. The diagnosis of feminizing testis or complete androgen insensitivity syndrome was established, leading to the decision to admit the patient for bilateral orchiectomy and sigmoid vaginoplasty. (Figure 3, 4)

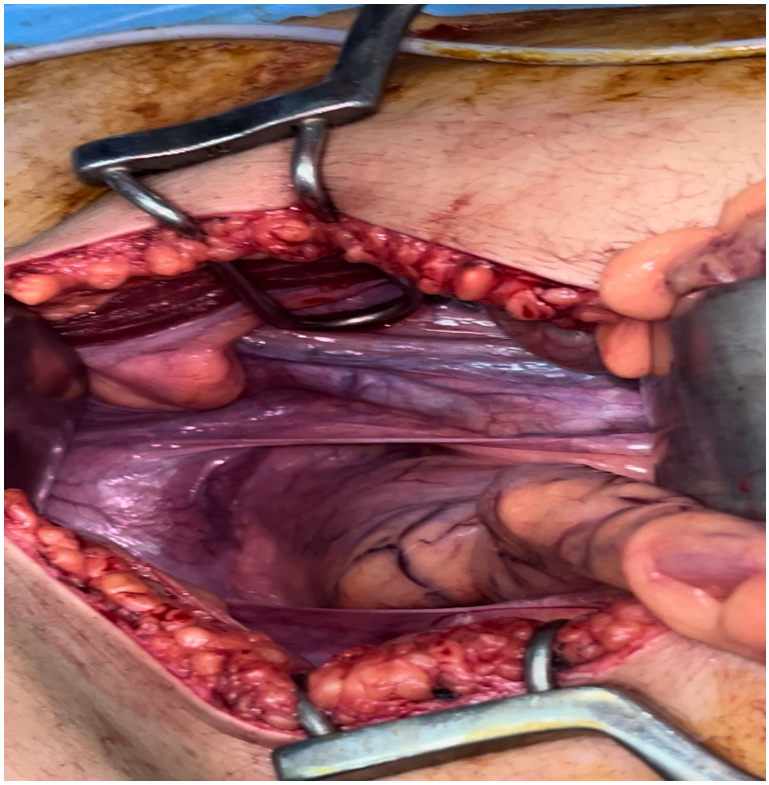

Figure 3: Intraoperative findings showing no vagina, uterus, adnexa, or prostate

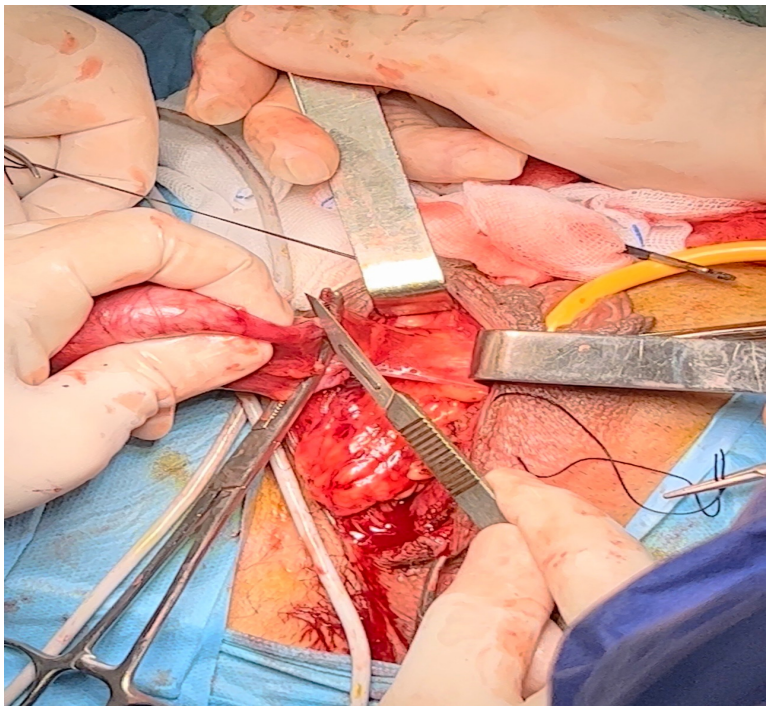

Figure 4: Gonadectomy

Case Report 2:

The second case features a 30-year-old woman admitted to the gynecology department due to primary amenorrhea. Clinical examination unveiled typical female physical characteristics, including well-developed breasts. However, her labia were underdeveloped, and there was an absence of axial hair in her pubic and groin regions. Gynecological examination revealed only a hymen and an underdeveloped vagina (2cm) while confirming the absence of a uterus.

Blood tests demonstrated normal levels of gonadotropins, such as FSH (6.64 mUI/mL), LH (26.65mIU/ml), progesterone (6.89 nmol/L), and estradiol within their respective limits.

However, testosterone levels were notably elevated at 58 ng/mL. Advanced screening for gene abnormalities related to the lack of Androgen receptor was considered, but unavailable.

Pelvic MRI revealed the absence of ovarian tissue and identified undescended bilateral testes within a hypoplastic uterine cavity. Subsequently, the patient underwent laparoscopic removal surgery conducted by specialists from the Urology Department. The procedure was successful, and the patient's postoperative condition notably improved rapidly, leading to her discharge within 24 hours. (Figure 4)

Histopathologic evaluation of seminiferous tubules from both removed bilateral testes samples confirmed severe Sertoli cell-only syndrome coupled with hyperplasia of Leydig cells. Moreover, the patient received female psychology counseling support to alleviate the anxiety associated with the initial revelation of her condition, along with estrogen replacement therapy to address hormonal imbalances. During counseling, it was explained to her that although genetically male, her body is not responding to male hormones and she has not developed typical masculine traits. While lacking complete female physical characteristics as well, she cannot be classified strictly as a female either. Despite this anatomical ambiguity however, she identifies psychologically as a woman due to being raised in such manner. She spoke openly about these issues during therapy sessions and received guidance on navigating the confusion surrounding her genetic makeup versus her physical attributes and mental image of herself. This anxiety could be eased by acknowledging the ways in which she might benefit from pursuing sexual relationships while realizing that giving birth would not be an option for her specifically due to neurological conditions caused by certain differences between the two biologically determined aspects of sexuality involved here: reproductive capacity vs. psychological gender identity.

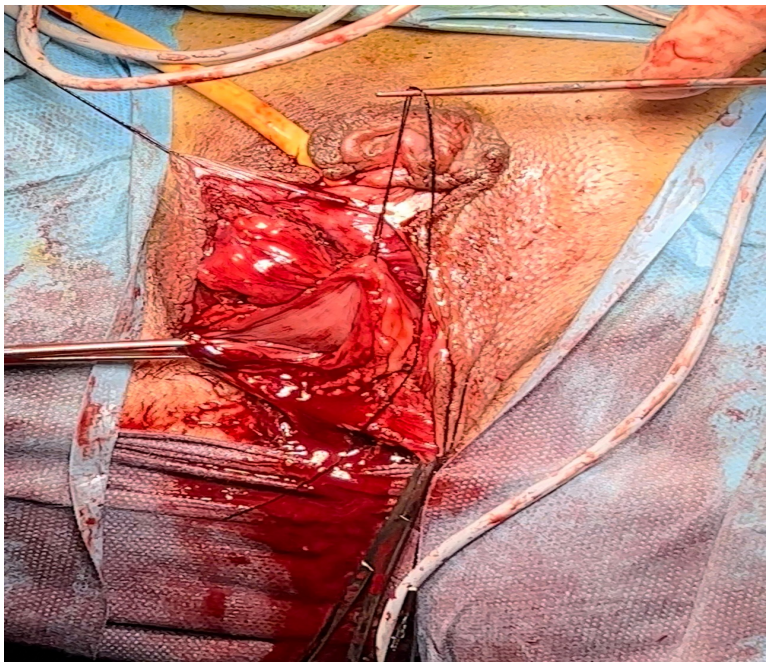

Figure 5: The edges of the sigmoid sewn to the pit of the vagina.

Following the intervention mentioned above, her depression showed improvement and she no longer experienced fleeting delusions. As a result, she began to feel better overall. After follow-up sessions, it was revealed that she is considering marriage with a male individual whom she presently has a non-physical relationship with.

Discussion

Vaginal agenesis is a health condition that affects about 1 in every 5,000 female live births and it's usually linked to Mayer-Rokitansky-Küster-Hauser syndrome (congenital aplasia of the uterus and upper 2/3 of the vagina, in women with normal development of secondary sexual characteristics and a normal 46, XX karyotype). It occurs as a result of incomplete development of the sinovaginal bulbs required for forming the vaginal plate [3]. The existence of this condition dates back to early 19th century researchers including Mayer and Rokitansky while Kuster and Hauser added more knowledge on associated skeletal or renal anomalies [3]. Diagnosis typically happens during adolescence when patients exhibit primary amenorrhea despite normal puberty. Owing to their typical external femininity, normal ovarian function alongside having a XX karyotype diagnosis may be delayed.

Another type of Disorder of Sex Development is Testicular Feminization Syndrome, which occurs when individuals with an XY karyotype display feminine physical traits and are typically raised as females [4, 5]. Common features include a superficial vagina, the lack of Mullerian duct structures and male gonads present within the body [6, 7].

Numerous techniques have been documented for vaginal reconstruction. The Frank procedure that doesn't require surgery is appropriate in cases where a pouch or dimple exists and involves slow mechanical widening utilizing hard dilators of increasing size to develop an inward folding of the vaginal dimple [8]. However, because this approach proves lengthy and typically elicits prolonged discomfort, patients tend not to follow through with it; consequently, practitioners don't advise using it on individuals who just possess skin depressions since they're less likely to recover successfully [9].

The McIndoe procedure and other surgical techniques involve placing a mold covered with either split-thickness or full-thickness skin graft, taken from the buttocks or labia majora, into the newly created neovaginal space. Postoperative vaginal dilation is necessary for successful results [10]. Other methods include using peritoneum from the Douglas pouch, amnion, oxidized regenerated cellulose fabric or muscle flaps; though these have been known to cause complications such as high incidence of vaginal stenosis and shortness in length leading to dyspareunia and dryness of vagina respectively [11- 15]. Dyspareunia occurrence depends directly on how long your new vagina is - there's 100% if it falls below six cm [16].

Originally developed in Italy, the Vecchietti procedure simplifies neovagina creation by utilizing a surgically placed traction system to accelerate dilation of the rudimentary vagina through use of a vaginal bead or "olive" [17, 18]. However, extended periods of vaginal dilatation and stenting with a mold at night are required for these methods which can negatively impact patients' mental health. Moreover, such techniques are inappropriate when treating pediatric populations [16].

Vaginoplasty has utilized bowel segments since 1904 when Baldwin initially introduced the procedure, which entailed isolating a U-shaped sigmoid colon and connecting it to the perineum. [19]. Nonetheless, due to its high mortality rate, this method was discontinued in the early 1970s. Nevertheless, Goligher documented an encouraging but restricted use of a pedicled portion of the sigmoid colon for constructing neovagina in the early 1980s [20]. In later years - specifically in 1987, Hanna detailed their experience with pure pediatric usage involving an ileal pouch for vaginoplasties [21].

Bowel vaginoplasty can involve a variety of bowel segments such as the colon, caecum or ileum; however, using the sigmoid colon is preferred due to numerous benefits [22]. These advantages include self-lubrication and reduced mucus production compared to small intestine use. Additionally, if employed prior to puberty there's growth potential with minimal risk for stenosis given its close proximity to the perineum. The sigmoid colon also has an easily mobilized vascular pedicle while eliminating molds or stents from necessary equipment for creation of a cosmetically pleasing neovagina that features appropriate length and natural lubrica-

tion via sigmoid vaginoplasty methods. [23, 24].

McQuillan and Grover's recent analysis revealed that bowel vaginoplasty remains the top- performed procedure, providing an average vaginal length of 12.87 cm [23]. Nonetheless, given that a significant majority of reviewed articles are either case reports or observational studies with limited sample sizes, determining the optimal treatment approach still requires further investigation.

While complications during sigmoid vaginoplasty are infrequently reported, we did not encounter any intraoperative or intestinal issues in our study. This can be attributed to the importance of thorough preoperative bowel preparation and skillful suturing techniques that contribute towards avoiding such occurrences. Our department has extensive expertise in managing various surgical procedures involving bowels.

After a sigmoid vaginoplasty procedure, patients often experience an excess of mucus production which is typically not worrisome and diminishes with time [24]. We had one patient in our series who required regular cleaning to manage excessive mucus produced during the perioperative period. However, this symptom gradually decreased over time.

Although mucosal or full neovaginal prolapse has been reported in some instances, with a prevalence of up to 14%, we did not observe this complication thanks to our consistent practice of sigmoidopexy. This technique involves attaching the proximal segment of the sigmoid colon to the sacral promontory. The incidence rates for introital stenosis following sigmoid vaginoplasty vary between 8.1% and 19.3% [25,26]. To minimize the risk of anastomotic site stricture, it is necessary to select a well-vascularized bowel segment that is long enough without tension when mobilizing it towards perineal region while creating sufficient space between bladder and rectum as well as ensuring meticulous hymenal anastomosis [27].

Whether regular vaginal dilatation is required after sigmoid vaginoplasty continues to be a controversial issue. Our study revealed that just one patient who was not sexually active experienced stenosis and needed consecutive dilatations. Therefore, while we do not advocate it as mandatory, we propose consistent postoperative dilation (minimum once per week) of the neovagina until sexual activity commences.

None of our patients experienced any major postoperative or long-term complications in the series. However, it is worth noting that rare long-term complications should be taken into consideration. In some instances, ulcerative colitis has been observed in the neovagina.

Additionally, individuals with specific hereditary polyposis syndromes such as familial polyposis, Gardner syndrome and nonpolypous colon cancer may develop polyps or neoplasia in their diverted sigmoid colon [28]. There are also isolated cases of diversion colitis- an unusual idiopathic disorder which can occur after isolating a segment of the intestine from fecal streams [29] - but these conditions are exceedingly uncommon within India's patient population and may explain why they were absent among our study participants.

An equally important aspect of care is psychological support for the patient during and after the operation, as part of a multidisciplinary approach [30]. Emphasis must be placed on the discrepancies between chromosomal, gonadal and physical sex characteristics, and their potential consequences. People with complete androgen insensitivity syndrome show a long-term psychosexual outcome that indicates a tendency to develop typically feminine traits, such as acceptance of their feminine identity and display of behaviors considered typically feminine. In addition, they experience psychological well-being comparable to that of other women [31].

Psychological intervention could potentially address problems arising from changes in sex-related self-image, as well as problems related to desire, arousal and dyspareunia [32]. If vaginal penetration can be a problem, vaginal dilators or regular penetrative sex could alleviate any effects arising from hypothetical vaginas, and help the patient become increasingly aware of her vagina, and of her status as a woman.

The significance of molecular analysis and the discovery of unfamiliar blood and tissue biomarkers is growing in importance as they serve as essential diagnostic, prognostic, or predictive methods in treating tumors that emerge from different sites such as genito-urinary and gynecological areas. Also, a multidisciplinary management of the patient is necessary including gynecologists, urologists, endocrinologists, and psychiatrists.

Conclusion

Sigmoid vaginoplasty combined with orchidectomy represents a comprehensive surgical approach offering a promising solution for individuals with AIS who seek to address their distinctive anatomical concerns. Current research underscores its potential to deliver both functional and psychologically advantageous outcomes, significantly enhancing the quality of life for AIS patients. Nonetheless, ongoing research and the collaboration of medical and psychological professionals are essential to continually refine surgical techniques and advance patient care in this specialized field.

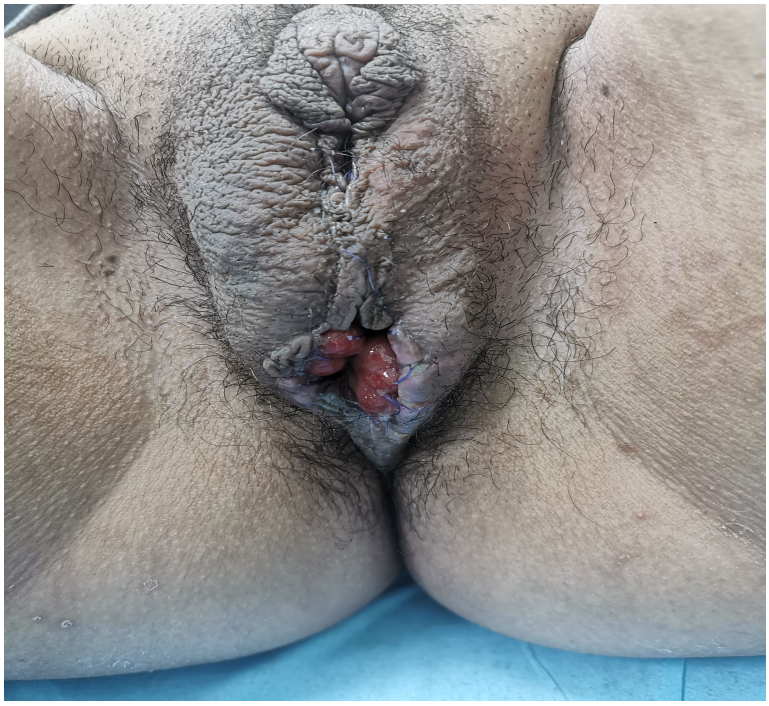

Figure 6: Post-operative result after 2 weeks

Declarations

Conflicts of Interest

The authors declare that they have no competing interests

Sources of Funding

There are no funding sources to be declared

Ethical Approval

Ethics approval has been obtained to proceed with the current study.

Ethical approval for this study (Ethical Committee N°09-24) was provided by the Ethical Committee Ibn University Hospitals, Rabat Morocco on 19 January 2024

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor- in-Chief of this journal

Guarantor of Submission

The corresponding author is the guarantor of submission

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Availability of Data and Materials

Supporting material is available if further analysis is needed

Provenance and Peer Review

Not commissioned, externally peer-reviewed

Scare Guidelines

The work has been reported in line with the SCARE criteria [33]

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