




A review of pediatric infravesical obstructions

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The term infravesical obstruction is used for functional or anatomical disorders that are located distal to the bladder neck and cause disruption or interruption of urinary flow. Among these pathologies, the most common cause of urinary system obstruction in male newborns is the posterior urethral valve. In this study, it was aimed to review the diseases that may cause infravesical obstruction in children.

Keywords: Child, infravesical obstruction, pediatric, posterior urethral valv, urethral stenosis.

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Cite this article as: Cansaran S, Ayvaz OD, Celayir A. A review of pediatric infravesical obstructions. Zeynep Kamil Med J 2024;55(1):1–7.

Received: January 05, 2023 **Accepted:** March 27, 2023 **Online:** February 12, 2024

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Zeynep Kamil Medical Journal published by Kare Publishing. Zeynep Kamil Tıp Dergisi, Kare Yayıncılık tarafından basılmıştır.

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INTRODUCTION

Infravesical obstruction is a term used for anatomical or functional pathologies distal to the bladder neck that affects and interrupts urine flow in different ways. Since infravesical obstructions can affect the entire lower and upper urinary tract, the morbidity is high, and the management of these patients is often quite difficult.

When infravesical obstruction is mentioned, the first pathology that comes to mind is the posterior urethral valve (PUV), which is the most common cause of urinary tract obstruction in male newborns. Urethral prolapse, meatal stenosis, megalourethra, urethral duplication, congenital urethral fistula, urethral stricture/stenosis, urethral atresia, urethral diverticula/anterior urethral valve, and cystic Cowper's gland duct are other less common pathologies that may cause infravesical obstruction.

PUV

PUV is the most common etiology of urinary tract obstructions in male newborns. Due to the valve, obstruction develops in the membranous folds of the posterior urethra. Its incidence is 1 in 4000–8000 pregnancies.^[1] In children, it is also the most common cause of chronic kidney disease (CKD) caused by urinary tract obstruction.^[2]

Pathogenesis

It is thought to originate from the persistent obstructive urogenital membrane. The mechanism is not clear. It is assumed that disruption of the normal embryological development of the urethra at 9–14th weeks produces obstructive membranous folds in the posterior urethral lumen. The most accepted embryological mechanisms are persistent urogenital membrane formation as a result of abnormal canalisation of the urethra, overgrowth of the urethrovaginal folds, and abnormal integration of the Wolffian duct into the posterior urethra.^[3]

Classification

In the traditional classification made by Young et al.^[4] in 1919 according to autopsy results, type 1 valve is a protrusion that emerges from the verumontanum and connects to the anterior urethra in the form of two leaflets and is the most common. Type 2 valve is no longer considered as a PUV and is assumed to be a dissection artifact. It has been reported that it extends vertically from the verumontanum to the internal sphincter and bladder neck. The type 3 valve is in the form of a diaphragm with a central perforation distal to the verumontanum, it has been reported that it may be a result of urethral catheterization.

In the 1990s, with a review after radiological and cystoscopic studies, it was suggested that PUV is a singular pathological condition in an oblique membrane associated with the verumontanum, termed the congenital obstructive posterior urethral membrane (COPUM).^[5] Although this approach has not been widely adopted in clinical practice, it has been possible to distinguish between COPUM and “Cobb's collar,” which is defined as a congenital urethral stricture caused by a transverse membrane in the anterior urethra and not associated with verumontanum.^[6]

Clinical Presentation

In developed countries, 1/3–1/2 of patients with PUV are diagnosed with prenatal ultrasound (US).^[1] Those diagnosed postnatally are usually newborns or young infants with abdominal distention, urinary tract symptoms, or lung hypoplasia associated respiratory distress.

Prenatal US shows bilateral hydronephrosis (HN), dilated bladder, and posterior urethra (keyhole sign) in male fetus.^[7,8] The bladder wall, which normally does not exceed 3 mm, may thicken. In severe cases, oligohydramnios is present. Urinary acid or perinephric urinoma may be seen as a result of fornix or calyx perforation caused by increased pressure. A large bladder diverticula or patent urachus may develop. Increased renal echogenicity and cortical cysts supporting renal dysplasia may also accompany the clinical condition.

An increased risk of perinatal mortality and postnatal CKD can be mentioned if PUV markers are present before the 24th week of gestation in the US, or there is severe bilateral HN with oligohydramnios, or there are findings compatible with renal dysplasia.^[9] The presence of oligohydramnios in the second trimester is associated with 90–95% perinatal mortality.^[10]

Patients who cannot be detected prenatally are usually diagnosed in the newborn or early infancy period. About half of the patients present with urinary tract infection.^[11] Respiratory distress due to pulmonary hypoplasia is one of the most common reasons for presentation in newborns with PUV. Abdominal distention due to excessively distended bladder or urinary acid, and difficulty urinating or weak urine flow are other symptoms of PUV in newborns.^[12] Growth retardation, urosepsis, weak urine flow, and straining or grunting during urination are more common in infants. Older children are more likely to present with urinary tract infection, day and night incontinence, and other signs of voiding dysfunction (increased voiding frequency, straining to void, weak urine flow, and large urine volume at each voiding).^[13]

Urologic Outcomes

CKD is commonly associated with PUV due to renal dysplasia and/or acquired renal damage due to infection, or problems with poor bladder function. About 15–20% of patients with PUV progress to end-stage renal disease (ESRD).^[14] In severe cases, renal replacement therapy (RRT) may be required in addition to medical management of renal failure complications due to infancy ESRD.

Renal dysplasia is common in patients with PUV, and parenchymal changes supporting renal dysplasia prenatally are seen in approximately 60% of infants diagnosed with PUV in the same period.^[14] High serum creatinine levels, which indicate impaired kidney function due to renal dysplasia, may be seen despite bladder decompression.^[15] The higher voiding pressures that infants may have, even after valve ablation, are not necessarily associated with impaired kidney function.^[16]

The relationship between renal dysplasia and PUV has not been explained yet. Possible mechanisms are described as extensive developmental damage resulting in renal dysplasia and PUV and a causal relationship between defective nephron development and back pressure from PUV's bladder outlet obstruction. The second theory is generally associated with the “pop off” mechanism. In this case, the pressure is reduced by formations and processes such

as bladder diverticulum, urinary acid due to extravasation, perirenal urinoma^[17] and massive reflux on one side, non-functional kidney on the same side, and normal development on the contralateral side.^[18]

Vesicoureteral reflux (VUR) is seen in approximately 1/3–1/2 of patients with PUV (N). It is in the form of reflux secondary to the closure defect of the ureterovesical junction caused by increased intravesical pressure. VUR resolves in at least one-third of patients with removal of the obstruction. In particular, bilateral involvement is a risk factor for poor kidney function and ESRD.^[15] In a study of 197 patients with PUV, VUR was detected in 127 cases, 73 of which were bilateral.^[19] Serum creatinine levels were found to be higher in patients with VUR at the time of PUV diagnosis and at 6th/12th months postoperatively following ablation. ESRD was seen in 25% of patients with bilateral VUR, 7% of patients with unilateral VUR, and 4% of patients without VUR.

Muscular hypertrophy and collagen deposition occur in the bladder wall due to outlet obstruction. Thickened bladder wall or trabeculations/diverticulum are the most common findings on US. These changes lead to uncontrollable bladder contractions (overactive bladder) and a decrease in compliance. Urodynamic (UD) studies typically reveal a bladder with low capacity, high filling pressure, and decreased compliance.^[15]

Diagnosis and Imaging

Probable diagnosis is made by voiding cystourethrography (VCUG) showing distinctive signs of dilated and elongated posterior urethra during the voiding phase. Based on the VCUG findings, cystoscopy is performed to confirm the diagnosis and for PUV ablation.

Renal and bladder US is performed to measure the degree of HN, to assess renal parenchymal thickness and corticomedullary differentiation. The bladder wall may appear thickened and is best evaluated when the bladder is full. If placement of the urethral catheter has been difficult, US is an excellent method for assessing the localization of the catheter.

Radionuclide scans are used to identify renal parenchymal anomalies and evaluate the degree of obstruction. Dimercaptosuccinic acid scintigraphy is the most useful method for detecting focal renal parenchymal abnormalities and evaluating differentiated function between the two kidneys. After intravenous injection, it is taken up by the proximal tubular cells and little is secreted into the urine. Thus, it accumulates in the tubules for several hours and provides a static image. Dynamic scanning with diethylenetriamine pentaacetate and mercaptoacetyl triglycine (MAG-3) evaluates the excretory function of the kidney. It is excreted into the lumen and then into the bladder by glomerular filtration and proximal tubule secretion. MAG-3 is the first choice for obstructions, even in neonates with immature glomerular filtration. The time for the radionuclide to drain from the renal pelvis is measured and the renal/excretory function of each kidney is evaluated.

Urethral agenesis or stricture/stenosis, megalourethra, megacystis, and microcolon syndrome should be kept in mind in the differential diagnosis of PUV.

Prenatal Intervention

Although the prenatal detection of PUV and the development of surgical equipments have increased the rate of prenatal surgery, still most patients are treated soon after birth. Prenatal intervention was first

applied in 1981 for obstructive uropathy.^[20] Urinary drainage through vesicoamniotic shunt aims to prevent lung hypoplasia by correcting oligohydramnios, and to improve long-term kidney function by reducing back pressure and nephron damage. However, proven long-term renal recovery has not been demonstrated, and there is an increased risk of fetal and maternal morbidity.

In a study conducted between 1988 and 1991, fetal intervention criteria included normal karyotype, the presence of appropriate levels of urinary electrolytes, and the absence of cystic renal disease on US.^[21] Thirty-six fetuses underwent surgical intervention, including 14 with PUV at a mean gestational age of 22.5 weeks. Vesicoamniotic shunt was performed in 9 patients, PUV ablation in two patients, bladder marsupialization in two patients, and ureterostomy in one patient. Five infants born prematurely and had respiratory failure died. One pregnancy was terminated due to evidence of shunt failure and findings indicative of poor lung/renal development and function. Eight patients survived with a mean follow-up of 11.6 years. Two of these had undergone renal transplantation due to CKD and one was waiting for donation. Similar results were seen in study conducted in 2016.^[22]

There is significant morbidity and mortality rates of prenatal intervention for PUV. Prenatal intervention should be considered only in fetuses with a high risk of in utero or neonatal death due to severe oligohydramnios in the mid-trimester and who have both a normal karyotype and good indicators of renal function based on fetal urinary evaluation. For the evaluation of fetal urine, the first urine obtained by US-guided catheterization is discarded and a sample is made from the urine collected in the second drainage. A favorable urine sample indicating good renal function has a urine sodium of <100 mmol/L, chloride <90 mmol/L, osmolality <210 mOsm/L, and a beta-2 microglobulin <6 mg/L.^[23] Due to the high maternal and fetal risk, surgical intervention should only be performed in fetal treatment centers with significant experience and expertise.

Postnatal Management

In the presence of suspected or diagnosed PUV, the first postnatal treatment is stabilization of the patient and drainage of the urinary tract. Correction of electrolyte abnormalities (especially hyperkalemia), treatment of respiratory distress and urosepsis, and monitoring of kidney functions at all stages constitute medical management. Temporary drainage of the urinary tract after patient stabilization is usually provided by a catheter inserted through the urethra into the bladder. In some centers, direct PUV ablation is performed.^[14] A soft feeding tube is a better option than a Foley catheter due to its larger internal diameter and thus better drainage (No.8 Fr or larger according to the structure of the urethra). The balloon of the Foley catheter can occlude the ureteral orifices, especially when overinflated. The catheter may bend in the wide posterior urethra and it should be checked with US when necessary. If the catheter cannot be placed, cystoscopy should be considered promptly.

After drainage is established, there may be a high loss of water and electrolytes in the urine due to tubular dysfunction.^[24] Tubular dysfunction occurs because urine cannot be concentrated and electrolytes cannot be absorbed. Furthermore, some patients with PUV may develop type IV renal tubular acidosis due to urinary obstruction. As a result, strict monitoring of serum electrolytes including bicarbon-

ate and fluid status should be performed, and fluid and electrolyte replacement therapies should be administered when necessary.

Cystoscopy and Other Procedures

Cystoscopy enables the diagnosis to be confirmed by direct demonstration of PUV. Primary ablation is the first choice and because it is less invasive, it contributes to the preservation of bladder function and reduces the need for subsequent surgery.^[14] The timing of intervention depends on the infant's general health and problems with anesthesia and is determined by whether the size of the urethra is suitable for the neonatal cystoscope. Valve ablation relieves urethral obstruction in most patients. With a decrease in bladder pressure after ablation, reflux will improve in one-third of patients with VUR.

Premature infants are a little more difficult to manage, urinary drainage can be achieved with smaller catheters (No.5 or 6 Fr). A temporarily placed feeding catheter is often sufficient for the urethra to become suitable for the cystoscope. An observational study of 126 male newborns revealed that progressive urethral catheter dilation increased the rate of successful primary ablation to 97%, compared to the 82% reported in the literature.^[25] PUV ablation can also be performed using a Fogarty catheter under direct observation with a cystoscope in premature newborns.^[26] Thus, the need for long-term catheter usage or vesicostomy is avoided. However, experience is the limiting factor for this procedure.

If valve ablation cannot be performed for technical reasons, vesicostomy is the next choice as it allows urinary drainage, reduces urinary system pressure, and is a reversible procedure. It allows cyclic filling-emptying of the bladder at low pressure. It is used in extremely premature infants where the valve cannot be demonstrated safely or ablation is not possible.

High diversions such as cutaneous ureterostomy and pyelostomy are rarely indicated and are not as good as vesicostomy. In the future, they require major upper system reconstruction. In addition, since they disrupt the bladder filling-emptying cycle, they affect bladder function more.^[27]

Post-procedure Management

Detecting and treating bladder dysfunction after a successful interventional procedure, monitoring renal function and, if necessary, coping with the outcomes of CKD are the two cornerstones of post-procedural management.

Bladder function should be evaluated with imaging and UD studies. The use of clean intermittent catheterization (CIC) and anticholinergic agents reduces bladder pressures in patients with severe bladder dysfunction (low capacity, high filling pressure, and low compliance). It can reduce that the rate of VUR still present after ablation. Continued HN and/or hydroureteronephrosis (HUN) after PUV ablation may be a reflection of abnormal bladder function. Treatments to improve bladder compliance and provide complete emptying (CIC and/or anticholinergic agents) and UD studies are indicated in these patients.^[28] Ensuring nocturnal bladder emptying with an indwelling catheter at night may be beneficial in patients with persistent HUN. In one study, successful results were obtained when applied to 18 male patients with persistent HUN after successful PUV ablation.^[29]

In a series of 119 patients, almost all of whom were treated with early bladder drainage and primary PUV ablation, one-third of the cases had severe bladder dysfunction requiring CIC.^[15] In another study, 27 of 65 patients had bladder dysfunction and only 3 required CIC. Continence was achieved in 42 of 55 patients who were toilet trained.^[14] In both studies, anticholinergic and alpha-blocker drugs were used according to the UD results. In a small case series of 18 patients with high voiding pressures and/or low bladder capacity, early administration of oxybutynin was associated with improved bladder function on UD tests before toilet training.^[30]

Ongoing monitoring of kidney functions is important in patients with PUV since the risk of CKD is high in the post-operative period. Management in patients with CKD is directed toward its associated complications. Patients with CKD may have urinary concentration disorder that causes excessive urinary losses. Exceeding the carrying capacity of the urinary tract can result in an enlarged/poorly functioning bladder and permanent HN. A nocturnal indwelling catheter may be effective for complete decompression of the urinary tract.^[31]

Outcome

Ten-year survival in patients diagnosed in the 1st year of life is over 90%. In a retrospective review of the Pediatric Health Information System database covering the years 1992 and 2006, it was determined that 34 of 685 pediatric patients diagnosed and treated with PUV within 1 year of age died. More than half of these deaths occurred during the first hospitalization and primarily due to pulmonary hypoplasia.^[32] The probability of 10-year survival in this cohort is 94%.

Despite prenatal diagnosis and early intervention, a significant number of PUV patients develop ESRD.^[32,33] In a multicenter study of 274 patients treated within the first 90 days of life, 42 (15%) patients received RRT (dialysis or renal transplantation) during a median follow-up of 6.2 years. Multivariate analysis showed that the only independent risk factor was the serum nadir creatinine level (SNC1) in the 1st year of life.^[33] In this study, an increased level of SNC1 increases the risk of RRT. The risk of progression to RRT up to the age of 10 was 0% for those with an SNC1 level <0.4 mg/dL, and 100% for those with an SNC1 level higher than 1 mg/dL.

Although it is thought that patients presenting later in life may have milder disease, it remains unclear whether a later presentation is associated with a better outcome. In a series of 315 patients, the rate of developing CKD in prenatally diagnosed cases (19%) after a median 5.5-year follow-up period was found to be lower than in cases diagnosed postnatally (40%).^[34] The mean serum creatinine levels for the two groups were similar at the time of diagnosis, but were lower in cases with prenatal diagnosis at the end of follow-up (0.96 vs. 1.75 mg/dL). Conversely, a small case series reported that patients presenting before the age of one were more likely to have poor renal outcomes.^[35] These results suggest that a significant number of patients with PUV will have renal failure, some will progress to ESRD and will require RRT. Persistently elevated serum creatinine level after removal of the obstruction is a risk factor for ESRD.^[15] The effect of the presentation age on the risk of renal failure remains unclear.

The survival of patients with PUV after renal transplantation is the same as in patients with renal transplantation for other primary causes of ESRD. In a study in which 418 pediatric cases with 8-year fol-

low-up were reported, there was no difference in the rate of allograft failure after renal transplantation among the patients with ESRD due to PUV or other causes.^[36]

After PUV ablation, patients are at risk of bladder dysfunction and may require CIC.^[33] There may be delays in achieving day and night urinary continence. Lower urinary tract symptoms are commonly seen in adult patients with PUV. In one study, adult patients with PUV (mean age 38.5 years) were more likely to have one or more moderate/severe lower urinary tract symptoms than their age and sex-matched controls (15.8% vs. 32.4%).^[37] Symptoms have been reported as mild hesitation, weak flow, incomplete voiding, spotting, and urge/stress incontinence.

URETHRAL PROLAPSE

It is a pathological condition that occurs in girls at an average age of 5 years and is more common in Africans. Dysuria, blood stains on underwear, and a ring of swollen, purplish, and prolapsed tissue in the meatal region are the most common symptoms and signs.

Sitz bath, antibiotic, or estrogen creams applied several times a day are usually sufficient in the treatment of mild prolapses. Placement of a temporary urethral catheter may be required. In persistent cases, excision of the prolapsed tissue and reanastomosis of the skin margins are curative. Other treatment options are simple reduction under general anesthesia or ligation of the prolapsed urethral epithelium over a Foley catheter.^[38] It should be noted that the ligation procedure performed over the catheter may cause post-operative pain.

MEATAL STENOSIS

Meatal stenosis is a pathological condition that is seen in males following circumcision and is thought to be caused by friction to the diaper or irritation.^[39] Meatal stenosis, which may occur for unspecified reasons, is always on the ventral side and causes the urine to divert to the dorsal side. Evaluation of voiding together with physical examination is important for diagnosis. If the urine flow is well calibrated or there is no dorsal deflection, no treatment is required. Rarely, dysuria or bleeding may occur with tearing of the web during voiding. Occasionally, inflammation around the meatus responds well to steroids (betamethasone 0.05%).

The surgical treatment of meatal stenosis includes simple meatotomy or sutured meatoplasty. Meatotomy can be performed in selected patients using topical local anesthetic agents (after 1 h with covering gauze) and oral midazolam.^[39] When the glans anesthesia is provided, the area to be cut is crushed with a hemostat clamp for 1 min, then it is cut to half the distance to the corona. The patient is given topical ointment treatment, in which he will apply several times a day for 2–3 weeks. Meatoplasty is similar to meatotomy, but in this technique, a thin suture is used to evert the urethral mucosa and prevent restenosis. Due to the sensitive suture technique, application under general anesthesia is required. Reoperation rates are less in meatoplasty (0.2% vs 3.5%), but office meatotomy is usually the first surgical treatment option due to its low restenosis rate and applicability with local anesthesia.^[40] Imaging studies and cystoscopy are not necessary to evaluate meatal stenosis.

MEGALOURETHRA

It is a rare genital anomaly, and the penis is deformed and elongated. It can be fusiform or scaphoid, and there are embryological and visual differences between these two forms. It is more common in patients with Prune-Belly syndrome and has been reported to be associated with VATER syndrome (vertebral defects, anorectal atresia, tracheoesophageal fistula, and renal dysplasia).^[41]

The scaphoid megalourethra is the most common form with a more benign character. There is an inability of the spongiosal tissue to form the urethra. Fusiform megalourethra is the more severe form. There is an inability of the penile mesoderm to form the spongiosal tissue or the corpus cavernosum. It is more associated with Prune-Belly syndrome, stillbirths, and cloacal anomalies.^[42]

Disease-related urological anomalies are megacystis, VUR, bladder diverticulum, and renal dysplasia.^[42] Therefore, upper system evaluation is indicated in all cases. Megalourethra repair is based on hypospadias techniques aimed at restoring the urethra to normal sizes.

URETHRAL DUPLICATION

It can be seen in various forms and is generally classified as dorsal or ventral to the normal urethra. Duplications may be complete or, more often, incomplete. Rarely seen side-by-side duplications are usually associated with the duplicated phallus and bladder. The urethra close to the rectum has a more normal spongiosal structure and sphincter mechanism and is more functional. The dorsal urethra is usually small and weak, also in an epispadiac position and associated with the dorsal chordee.^[43]

Effman classification is used to define duplication types. Partial duplications along the penile urethra are called Y-type, but when the duplicated opening is in the perineum, it is called H-type duplication.^[44]

Urethral duplication treatment is patient-specific. When only a small septum is present, endoscopic division of the septum may be successful. In more severe duplications, hypospadias techniques have traditionally been used to lengthen the ventral urethra to the glans penis. Progressive dilatation is recommended for functionalization of the dorsal urethral canal.

CONGENITAL URETHRAL FISTULA

Incomplete development of spongiosal tissue triggers diverticulum formation in the antenatal period. Congenital fistula occurs in the anterior urethra after urethral rupture due to the weak structure of the diverticulum.^[45] It is a rare entity and reconstructive treatment is difficult due to the absence of surrounding spongiosal tissue.

URETHRAL STRICTURE/STENOSIS

Most urethral strictures are acquired. Trauma, inflammatory processes and instrumentation are the most common causes. Congenital urethral stenosis is rare and mostly focal. The stenoses are usually at the junction of the bulbar urethra (genital fold) and the membranous urethra (urogenital sinus). Misalignment or incomplete drainage is implicated in the development of focal strictures.^[45]

Treatment options for both acquired and congenital urethral strictures include internal urethrotomy, resection and end-to-end anastomosis, and urethroplasty with flap or free graft.^[46]

URETHRAL ATRESIA

To be compatible with life, patent urachus must be present in the presence of urethral atresia. Reconstruction can be difficult, so a catheterizable stoma appears to be the best option after vesicostomy.^[47]

URETHRAL DIVERTICULA/ANTERIOR URETHRAL VALV

It can occur when the spongiosum is absent or thin. The distal portion of the diverticulum functionally acts as an “anterior urethral valve” and antegradely blocks the flow of urine. Urethral compression increases further as the diverticulum fills during voiding. Proximal dilatation occurs as a result of the valve effect.^[48] In some cases, valve effect does not occur, but a narrow-necked diverticula can cause urinary stasis and urethral infection. Urethrogram or cystoscopy is helpful in diagnosis.

Treatment options include cutting the distal part of the diverticulum causing the valve effect by cystoscopy and repairing the urethral defect after open excision of the diverticulum.^[49]

CYSTIC COWPER’S GLAND DUCT

It is a pair of small glands located in the urogenital membrane and is analogous to the Bartholin glands in females. Its ducts open into the ventral wall proximal to the bulbar urethra after exiting the gland. When the ducts are occluded, filling defect and sometimes obstruction occur in the bulbar urethra.

The cystoscopic finding is a thin membrane over a fluid-filled cyst (syringocele).^[50] If contrast material enters during imaging, tubular ducts parallel to the bulbar urethra can be seen. Treatment of cowper gland cysts is cystoscopic unroofing, but treatment is not necessary if there are no clinical symptoms.

Statement

Author Contributions: Concept – AC, ODA, SC; Design – AC, SC; Supervision – AC, SC; Materials – AC, ODA, SC; Data Collection and/or Processing – SC; Analysis and/or Interpretation – AC, SC; Literature Search – SC; Writing – SC; Critical Reviews – AC, ODA, SC.

Conflict of Interest: The authors have no conflict of interest to declare.

Use of AI for Writing Assistance: Not declared.

Financial Disclosure: The authors declared that this study has received no financial support.

Peer-review: Externally peer-reviewed.

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