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Robot-assisted staged bilateral reno-lymphatic disconnection for massive idiopathic chyluria: A case report

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ABSTRACT

Chyluria, an abnormal lymphatic disorder, results in excessive abdominal lymph drainage into the urinary system, causing protein loss, nutritional deficiencies, and immune issues. Mainly linked to parasitic infections in developed countries, non-parasitic causes like trauma or tumors are rare. Typically appearing in adults with bilateral involvement, management options include conservative or surgical approaches. We present the case of a 13-year-old with congenital chyluria, treated with robot-assisted staged reno-lymphatic disconnection after failed interventional radiology. Bilateral scleroangiography followed, leading to persistently milky urine for a month. Finally, urine clarity improved, correlating with better urinalysis, emphasizing the need for a comprehensive, multi-disciplinary approach.

1. Introduction

Chyluria is a rare disorder characterized by the presence of chyle in the urine. The incidence of chyluria is unknown, and the literature evidence is limited to case reports and small series.[1]

It is usually diagnosed in adulthood, and the etiology is the parasitic infection by Wuchereria bancrofti (filariasis) in 95% of the cases found in tropical and subtropical areas. Non-parasitic causes are instead extremely rare and represent most of the diagnoses in developed countries. Trauma, surgery (i.e. partial nephrectomy, aortoiliac bypass), malignancy, radiation, pregnancy, renal or bladder lymphangioma, thoracic duct stenosis, and pregnancy have been reported as the non-parasitic causes of chyluria.[2] Eventually, idiopathic presentations are attributed to congenital or acquired anatomical abnormalities of the urinary lymphatic system. The prognosis of non-parasitic chyluria is usually very good.

The etiology is related to the internal composition of the lymphatic drainage of the kidney. It occurs in a trilaminar fashion: the first lamina lies within the renal parenchyma, the second at a subcapsular level, and the third within the perinephric fat. The lymphatics in the second and third lamina communicate freely with each other. The intrarenal lymphatics emerge as 4–7 trunks, which emerge at the renal hilum to join the 2nd and 3rd level lymphatics, eventually converging along the renal vessels to the lateral aortic nodes. Chyluria occurs after the rupture of lymphatic varices in the renal tubules. The lymphatic varices result from high endo-lymphatic pressure, usually due to obstruction or stenosis of the main lymphatic ducts.[3]

The disease is often asymptomatic; in 70% of the cases, the first sign is the appearance of "milky" urine. Polysymptomatic cases have been described in patients with dysuria, hematuria, urinary retention secondary to chylous clots, edema, hydroceles, hypoproteinemia, weight loss, and malnutrition.[4] Based on the symptoms, the frequency of the episodes, and the extent of the calyceal involvement, it is classified as mild, moderate, or severe. The ether test, also known as the lipuria test, is a diagnostic test used to confirm the presence of chyluria. It involves collecting a urine sample from the patient and adding a few drops of diethyl ether. If chyle is present in the urine, the ether will dissolve the fat content, causing the urine to become milky or turbid. This change in appearance indicates the presence of chyle, confirming the diagnosis of chyluria. Nevertheless, lymphangiography is the gold standard, although invasive and not routinely used.[5]

Conservative dietary therapy is the first-line treatment with a 70% success rate.[2] For non-responder patients, sclerotherapy is an option, but there are no common guidelines for its use. Evidence has already shown the feasibility of robotic surgery to treat persistent chyluria in an adult patient,[6] but no data are available regarding pediatric patients with

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congenital abnormalities.

Herein, we reported the case of a 13-year-old boy affected by severe chyluria secondary to a bilateral abnormal connection between the lymphatic and the urinary systems. The patient was treated with bilateral robot-assisted lymphatic disconnection.

This manuscript was prepared following the CARE guidelines (https://www.care-statement.org).

2. Case report

A 13-year-old Caucasian boy with a BMI of 17 and no relevant past medical history complained of persistent milky urine for three months in mid-2020, raising the suspicion of chyluria. After its confirmation and the exclusion of either parasitic infections or previous trauma, the patient was admitted and hospitalized, suspecting reno-lymphatic abnormal communication as the etiology. He underwent a contrast-enhanced thoraco-abdominal MRI in July 2020, showing lipoproteinaceous aggregates in the upper (>left) and lower urinary tract (Fig. 1A).

The patient developed chyluria-related immunodeficiency (hypogammaglobulinemia and lymphopenia); therefore, he received a long-term course of antibiotics, a hypolipidemic diet, and immunoglobulin replacement therapy.

The patient was initially followed up elsewhere. In September 2020, he underwent lymphangiography, which described a retrograde flow of contrast medium through the renal parenchymal lymphatic vessels bilaterally, the left renal pelvis, and the bladder (Fig. 1B). Therefore, bilateral scleroangiography was performed.

No further episodes were reported for five months until April 2021, when chyluria recurred. Left pedal lymphangiography showed bilateral renal parenchymal lymphatic vessels opacification due to lymphatic crossover channels (Fig. 2). The patient finally came to our observation in November 2021.

After the interventional radiological management failed, a robot-assisted transperitoneal left reno-lymphatic disconnection was performed in December 2021 with stripping of the kidney vascular pedicle components and the proximal ureter, plus perirenal fat excision. The surgery was uneventful. Blood loss was negligible. The patient was discharged after 4 days.

In February 2022, another MRI lymphangiography was performed. It showed right perirenal lymphatic vessel hypertrophy with upper urinary tract hyperintensity (a direct sign of abnormal communication), and left upper urinary tract hyperintensity (an indirect sign of abnormal communication) (Fig. 3A and B). Therefore, retrograde pyelography confirmed the MRI findings (Fig. 4A and B). Bilateral external ureteral

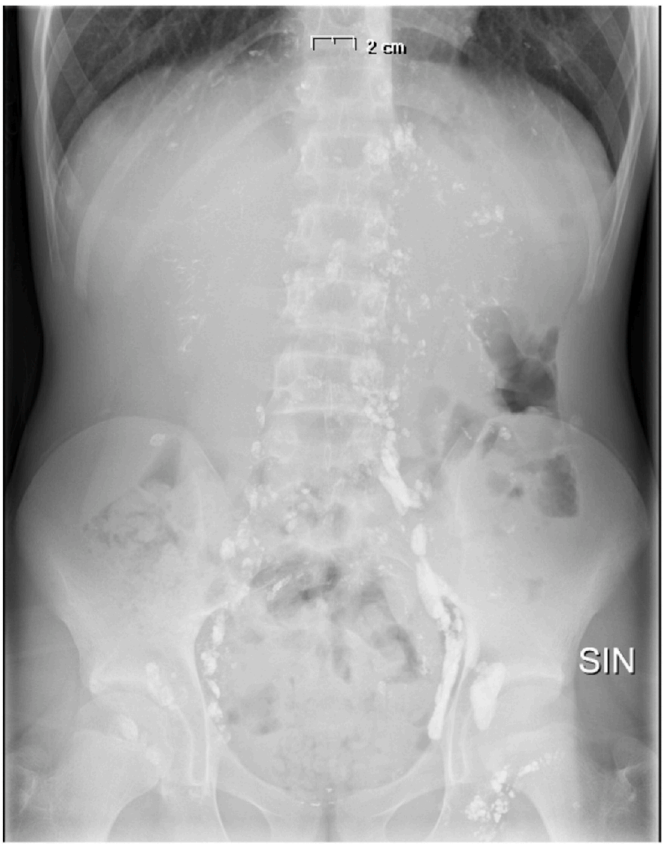

Fig. 2. Left pedal lymphangiography shows bilateral renal parenchymal lymphatic vessels opacification due to lymphatic crossover channels.

(mono-J) stents were placed to calculate the 24-h urine protein output, which was 2.13 g/24h on the right vs. 1.05 g/24h on the left.

In October 2022, the patient underwent robot-assisted transperitoneal right reno-lymphatic disconnection. Again, the surgery was uneventful. No chyluria was observed for 5 months after the second procedure. Subsequently, the urinary protein load gradually increased to the preoperative level (last evaluation = 14 g/24 h). A lymphography was repeated in July 2023, and a relapse of the disease was noted (Fig. 5A,B,C). Another bilateral scleroangiography was performed. The urine was persistently milky for the next month. Finally, a progressive

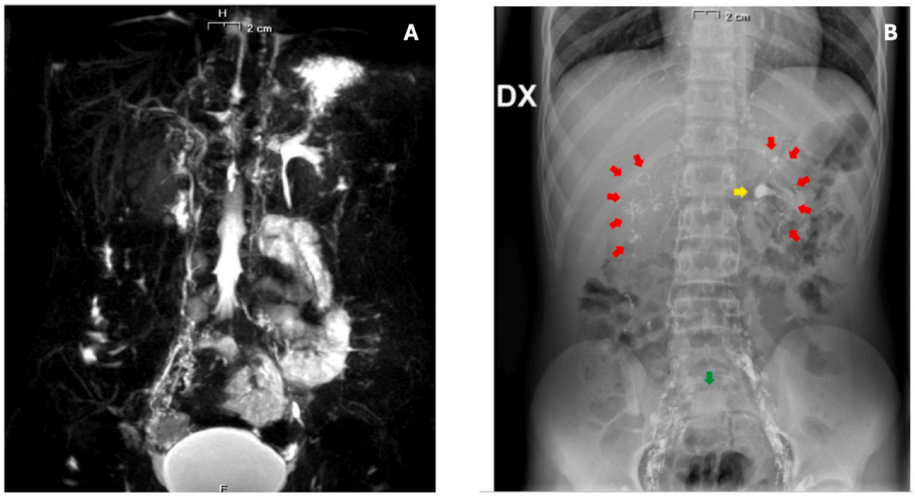

Fig. 1. a) Thoraco-abdominal-MRI shows the presence of voluminous lipo-proteinaceous aggregates in the bladder lumen; b) Lymphangiography shows bilateral renal parenchymal lymphatic vessels opacification (red arrow), left renal pelvis opacification (yellow arrow), and bladder opacification (green arrow).

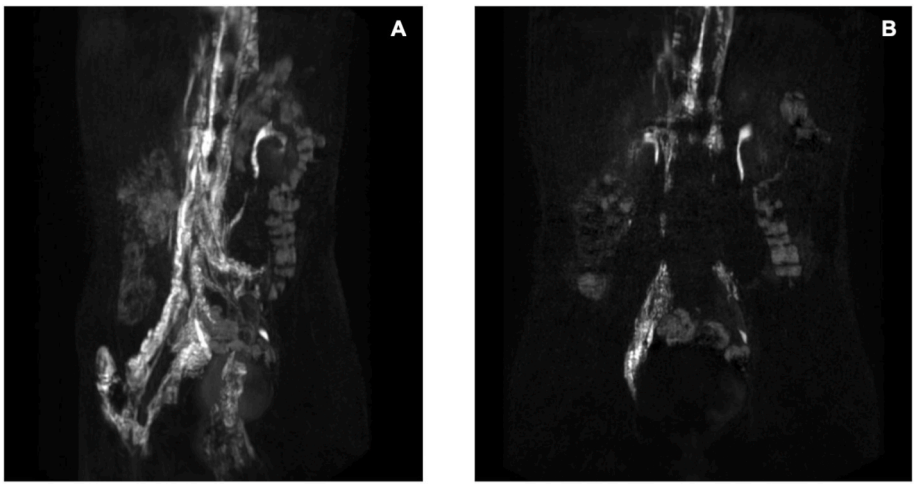

Fig. 3. MRI lymphangiography. It showed a) right perirenal lymphatic vessel hypertrophy with upper urinary tract hyperintensity (a direct sign of abnormal communication) and b) left upper urinary tract hyperintensity (an indirect sign of abnormal communication).

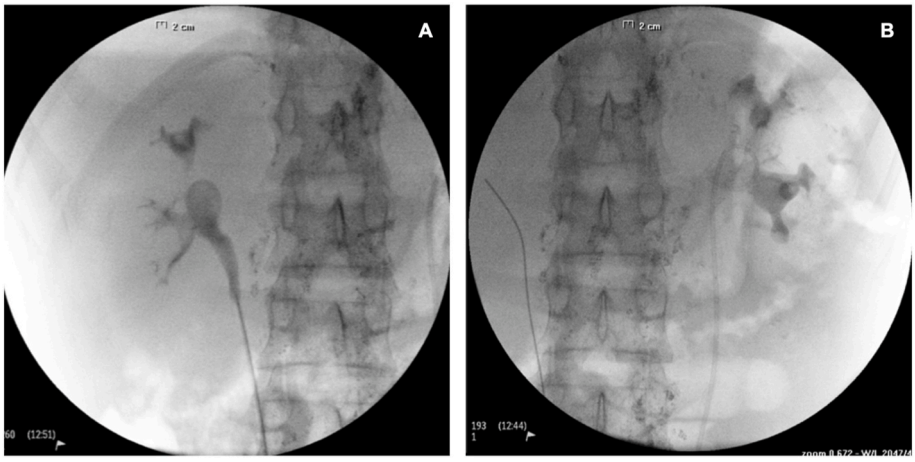

Fig. 4. a) Right retrograde-ureteropyelography shows early opacification of the hilar, retrocaval to iliac-obturator lymphatic pathways; b) Left retrograde-ureteropyelography shows early opacification of lymphatic and lymph node chains at the hilar, periaortic, and distally up to the iliac-obturator level.

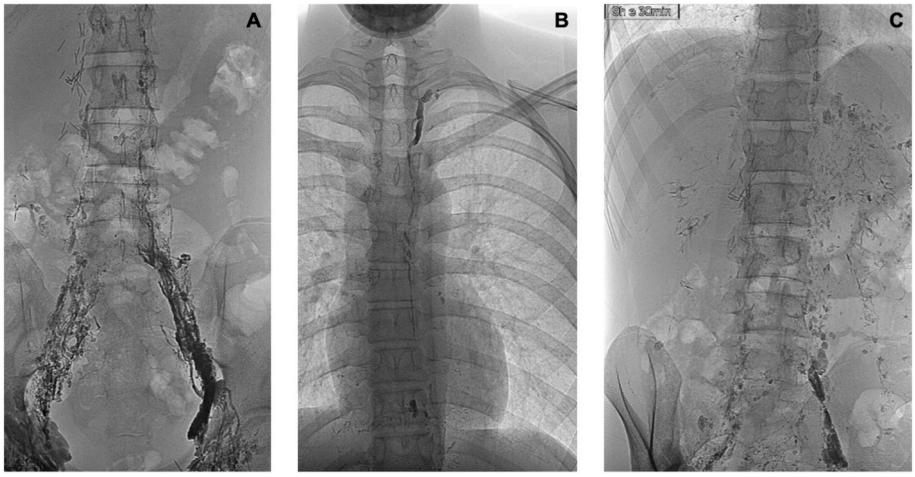

Fig. 5. a, b) Lymphangiography performed after surgery shows the immediate opacification of the iliac lymphatic vessels, followed by the abdominal lymphatic vessels and the thoracic ones up to the base of the neck; c) after 9 hours and 30 minutes, contrast medium is evident bilaterally in the pelvis and the renal calyces.

urine clarification was achieved, which was also associated with an improvement in the urinalysis (0.23g/24h) till the absence of proteins in the urine. The patient is still free from chyluria at eight months follow-up.

3. Discussion

We reported a rare case of bilateral massive chyluria in a pediatric patient, leading to acquired immunodeficiency due to protein loss in urine. The condition was due to an abnormal hypertrophy of the perirenal lymphatic vessels, with imaging showing the direct flow of the abdominal lymphatic stream to the excretory system.

For our young patient affected by severe chyluria, a non-invasive treatment was initially attempted, which lasted only a few months. Following the unsuccessful outcomes of conservative management, we opted for a surgical solution guided by the limited similar cases reported in the literature.

The robotic reno-lymphatic disconnection was first performed on the side where the phenomenon was more pronounced, achieving a partial relief of the proteinuria; due to the subsequent appearance of a massive lymphatic shift on the contralateral side, the procedure was repeated. However, again, the effect lasted only 5 months. In fact, the last lymphangiography performed in July 2023 showed an immediate opacification of the iliac lymphatic vessels, followed by the abdominal lymphatic vessels and, finally, the thoracic ones up to the base of the neck. Then, at 2, 4, and 9 hours, contrast medium was seen bilaterally in the pelvis and the renal calyces. Consequently, bilateral scleroangiography was repeated. Milky urine lasted for the next month, then progressive urine clarification was obtained, which was also associated with an improvement in the urinalysis. One of the last urinalyses showed a complete absence of proteins and mucus, usually present in the previous exams.

Given the rarity of the condition and the management adopted, we felt this case was worth discussing.

As mentioned, chyluria can be classified from mild to severe.[5] Depending on the severity, it is initially treated conservatively with dietary restrictions and supportive care, which in our patient consisted of regular immunoglobulin infusions. However, recurrence of chyluria after initial resolution has been reported in up to 80% of the patients treated conservatively. In severe chyluria, such measures are generally ineffective, so sclerotherapy or surgical treatment may be appropriate.[2] With sclerotherapy, the recurrence rate improves significantly to 13–41%. Nevertheless, the primary treatment failure rate with sclerotherapy ranges between 10% and 20%.[6] Recurrence of chyluria after surgery is usually due to incomplete stripping, reflux from the contralateral side, and reflux from the bladder. Methods to prevent recurrence include the use of an omental wrap or the use of enhanced visualization techniques such as magnifying loops or an operating microscope.

This highlights that there is still no standard treatment for chyluria.

The goal of the procedure is to seal and remove the abnormal reno-lymphatic communication, which results in increased pressure within the lymphatics and subsequent rupture within the urinary system. Robotics has been described as an effective treatment for this condition, but in an adult patient who had chyluria secondary to ureteral obstruction due to adhesions.[7] Again, Senthil Ganesh et al. described the same technique performed unilaterally, and via a laparoscopic approach in a 5-year-old child. In this case, the patient had the same management as ours and remained asymptomatic after the procedure.[8] On the contrary, Saebeom Hur et al. described a long history of chyluria in a 59-year-old patient who underwent 3 bilateral robotic/laparoscopic ureterolyses and skeletonizations with lymphatic stripping, which did not improve her symptoms.[9]

In our experience, we have found that severe chyluria cannot be effectively treated with reno-lymphatic disconnection alone. Without addressing the underlying anatomical condition causing abdominal lymphatic stagnation, new perirenal lymphatic vessels tend to replenish, resulting in a risk of relapse. The message we would like to emphasize with this case is that when chyluria has a congenital basis, it becomes a perilous disease that necessitates a multi-disciplinary, multi-modal treatment.

4. Conclusion

This case report emphasizes that when chyluria has a congenital basis, it should be approached from a multimodal perspective involving several professionals.

Statements

1) Informed consent was obtained from the patient.
2) All authors attest that they meet the current ICMJE criteria for Authorship.

CRediT authorship contribution statement

Sonia Costantino: Writing – original draft, Validation, Methodology, Data curation. **Francesca Montanaro:** Writing – original draft, Validation, Methodology, Data curation. **Alberto Bianchi:** Validation, Software, Resources, Data curation. **Greta Pettenuzzo:** Visualization, Validation, Data curation. **Riccardo Bertolo:** Writing – review & editing, Validation, Supervision, Project administration. **Maria Angela Cerruto:** Visualization, Validation. **Alessandro Veccia:** Writing – review & editing, Visualization, Validation, Resources, Data curation. **Alessandro Antonelli:** Writing – review & editing, Validation, Supervision.

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References

1. Mendu DR, Sternlicht H, Ramanathan LV, et al. Two cases of spontaneous remission of non-parasitic chyluria. *Clin Biochem*. 2017 Oct;50(15):886–888. https://doi.org/10.1016/j.clinbiochem.2017.05.002. Epub 2017 May 4.
2. Stainer V, Jones P, Juliebø SØ, Beck R, Hawary A. Chyluria: what does the clinician need to know? *Ther Adv Urol*. 2020 Jul 16;12, 1756287220940899. https://doi.org/10.1177/1756287220940899.
3. Gulati MS, Sharma R, Kapoor A, Berry M. Pelvi-calyceal cast formation following silver nitrate treatment for chyluria. *Australas Radiol*. 1999 Feb;43(1):102–103. https://doi.org/10.1046/j.1440-1673.1999.00604.x.
4. Singh DP, Sharma N. Chyluria: a clinical and diagnostic stepladder algorithm with review of literature. *Indian J Urol*. 2004;20(2):79.
5. Guttilla A, Beltrami P, Bettin L, et al. Chyluria: the state of the art. *Urologia*. 2017 Apr 28;84(2):65–70. https://doi.org/10.5301/uj.5000225. Epub 2017 Mar 27.
6. Dalela D. Issues in etiology and diagnosis making of chyluria. *Indian J Urol*. 2005;21(1):18–23.
7. Barman N, Palese M. Robotic surgery for treatment of chyluria. *J Robot Surg*. 2016 Mar;10(1):1–4. https://doi.org/10.1007/s11701-016-0560-5. Epub 2016 Feb 9.
8. Ganesh KS, Rao S, D'Cruz AJ. Successful laparoscopic management of unilateral chyluria in a 5 year old. *J Pediatr Urol*. 2007 Feb;3(1):63–65. https://doi.org/10.1016/j.jpurol.2006.02.003. Epub 2006 Apr 17.
9. Hur S, Gurevich A, Nadolski G, Itkin M. Lymphatic interventional treatment for chyluria via retrograde thoracic duct access. *J Vasc Intervent Radiol*. 2021 Jun;32(6):896–900. https://doi.org/10.1016/j.jvir.2021.03.410. Epub 2021 Mar 6.