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CASE REPORT OPEN ACCESS

Varicocelectomy Restores Spermatogenesis in Fontan-Associated Varicocele: A Two-Case Series

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ABSTRACT

Introduction: Fontan circulation may cause chronic venous hypertension and predispose patients to varicocele and infertility, but the clinical significance of surgical treatment remains unclear.

Case Presentation: We describe two patients with Fontan circulation who developed Fontan-associated varicocele (FAVC). A 42-year-old man presented with secondary infertility, bilateral varicocele, and progression to azoospermia after Fontan surgery. A 28-year-old man had asthenozoospermia, bilateral varicocele, and repeated failure of assisted reproductive technology. Both underwent bilateral microsurgical varicocelectomy with perioperative heparin bridging. Postoperative semen parameters improved in both patients, with sperm reappearing in the first case.

Conclusion: Varicocelectomy for FAVC may restore spermatogenesis in selected cases, but the risk of postoperative recurrence may be increased.

1 | Introduction

Fontan surgery is a widely used procedure for single-ventricle congenital heart disease (Figure 1) [1]. Despite improved survival, chronic venous congestion can cause long-term complications, including arrhythmia, heart failure, and liver cirrhosis [2, 3].

Chronic venous congestion due to Fontan physiology may lead to the development of an acquired varicocele, which we define as *Fontan-associated varicocele (FAVC)*.

2 | Case Presentation

2.1 | Preoperative Course

2.1.1 | Case 1

At age 37, the patient presented with testicular discomfort and was diagnosed with bilateral grade 3 varicocele. Initial semen analysis (SA) showed a sperm concentration of 15.2×10^6/mL, and he chose conservative management because of perioperative risk.

Keynote Message

- Fontan circulation can contribute to varicocele and infertility because of chronically elevated venous pressure.
- Despite possible recurrence and the need for careful perioperative management, microsurgical varicocelectomy should still be considered in selected patients.

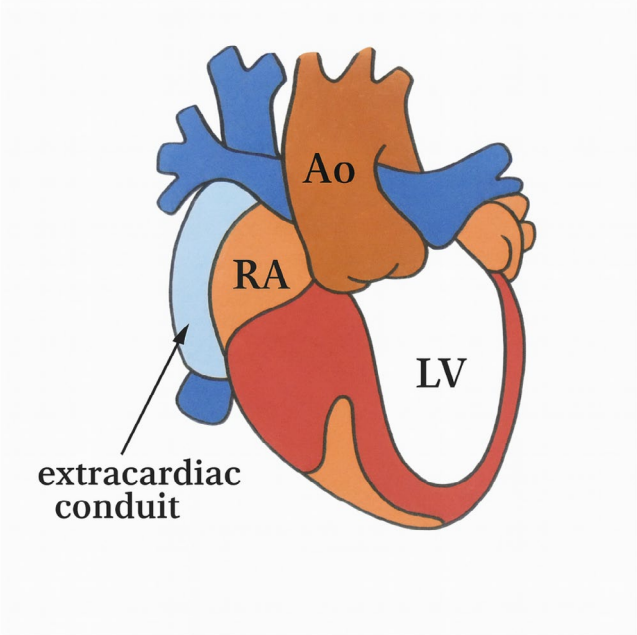

FIGURE 1 | Schematic of Fontan circulation.

At age 42, he underwent conversion from Bjork Fontan to total cavopulmonary connection (TCPC). After TCPC, central venous pressure (CVP) was 8–9 mmHg. Lower limb edema improved, although the varicocele persisted. Several months later, he presented again to our department with bilateral testicular pain and discomfort. SA showed cryptozoospermia, progressing to azoospermia 6 months after TCPC. Scrotal ultrasonography demonstrated bilateral grade 3 varicoceles with testicular volumes of 12 mL on the right and 10 mL on the left; intratesticular varicocele was not evident.

His history included Bjork Fontan surgery for tricuspid atresia, repeated surgeries for bilateral lower extremity varices, and chronic warfarin therapy. He had previously fathered one child naturally.

2.1.2 | Case 2

A 28-year-old man was referred to our department with a left grade 3 and right grade 1–2 varicocele, 2 years after undergoing TCPC. Initial SA showed preserved sperm concentration and motility.

Several months later, ultrasonography revealed bilateral testicular volumes of 18 mL and demonstrated left grade 3 and right grade 2 varicoceles. Intratesticular varicocele was not evident. SA then showed reduced total and progressive motility despite preserved sperm concentration.

His history included a right modified Blalock–Taussig shunt, conversion to TCPC, post-TCPC CVP of 10–11 mmHg, chronic warfarin therapy, and infertility for more than 2 years with repeated failed assisted reproductive technology (ART).

2.2 | Perioperative Course

Both patients underwent bilateral microsurgical subinguinal varicocelectomy under general anesthesia with perioperative heparin bridging therapy for continuous anticoagulation. Both patients developed grade 1 subcutaneous hemorrhage that resolved conservatively.

2.3 | Postoperative Course

Postoperatively, sperm reappeared in the ejaculate of Case 1, allowing him to proceed with intrauterine insemination with partner semen (Table 1). Semen parameters improved in both patients over time. In Case 1, sperm concentration and total sperm count continued to improve during follow-up despite recurrence of Grade 3 varicocele at 3 months. Serum FSH and testosterone levels remained stable throughout the follow-up period.

Preoperative ultrasonography revealed dilatation of pampiniform plexus veins in Case 1, and reversal of venous flow with marked enlargement during the Valsalva in Case 2 (Figure 2). Postoperative ultrasonography demonstrated a marked reduction in the diameter of the pampiniform plexus veins by 1 month after varicocelectomy. At 3 months, however, Case 1 developed a recurrence of Grade 3 varicocele, whereas Case 2 exhibited mild subclinical recurrence, characterized by dilatation of several veins. At the 6-month follow-up, neither patient had achieved pregnancy.

3 | Discussion

After TCPC, systemic oxygenation and cardiac output generally improve because of more efficient pulmonary circulation [2]. However, the circulation is achieved at the expense of chronically elevated central venous pressure, which results in persistent systemic venous congestion [4]. Consequently, organs drained by the systemic venous system—including the liver, kidneys, and pelvic organs—are continuously exposed to chronically elevated venous pressure, predisposing patients to complications such as hepatic congestion, ascites, peripheral edema, and varicocele [2–5]. Liver congestion occurs in most Fontan patients, and lower extremity venous reflux has been reported in about 50% of adults [5, 6].

In post-Fontan patients, ovarian dysfunction is associated with infertility and miscarriage [7]. However, there are no reports regarding the impact of Fontan physiology on male infertility. In these cases, systemic venous congestion may have contributed

TABLE 1 | Temporal changes in semen parameters before and after surgery.

	Case 1				Case 2			
SA	**Preoperative**	**POD 1 mo**	**POD 3 mo**	**POD 6 mo**	**Preoperative**	**POD 1 mo**	**POD 3 mo**	**POD 6 mo**
Volume (mL)	3.8	6.9	3.6	4.0	3.0	—	3.8	4.1
Concentration ($\times 10^6$ /mL)	0	3.3	9.6	16.2	30.0	—	45.0	56.0
Total count ($\times 10^6$)	0	22.8	34.6	64.8	90.0	—	171.9	229.6
Motility (%)	0	41	54	60	40	—	29	72
Progressive motility (%)	0	27	44	33	11	—	13	28
Abnormal morphology (%)	—	85–95	25–30	85–95	40–45	—	80–90	80–90
FSH (mIU/mL)	15.0	26.7	28.0	21.5	6.9	—	16.7	19.9
Testosterone (ng/mL)	9.2	8.6	7.4	6.4	11.2	—	5.2	7.3

Abbreviations: FSH, follicle-stimulating hormone; mo, month; POD, postoperative day; SA, semen analysis.

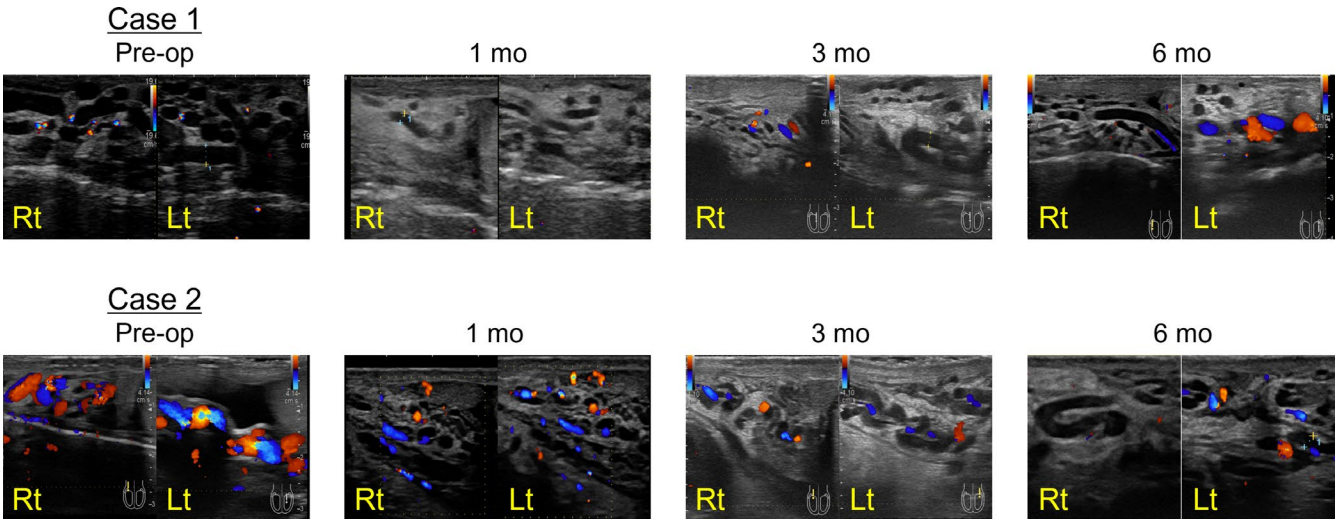

FIGURE 2 | Temporal changes in Doppler ultrasound findings of varicocele before and after surgery. Serial Doppler ultrasound images of the bilateral pampiniform plexus before surgery and at 1, 3, and 6 months postoperatively. Images were selected from comparable anatomical levels where venous dilatation was most prominent, with the patient standing during the Valsalva maneuver. Initial improvement at 1 month was followed by recurrent venous dilatation during follow-up. Scale bars are shown in each panel.

to the development of an acquired varicocele. In Case 1, SA deteriorated to azoospermia preoperatively but improved to within the normal range after varicocelectomy. To our knowledge, these are the first reported cases of microsurgical subinguinal varicocelectomy for FAVC.

Varicocele is a common cause of impaired spermatogenesis [8]. Proposed mechanisms include abnormal genetics, incompetent spermatic vein valves, increased venous pressure, and aberrant venous anatomy [9]. Few reports have described an association between elevated central venous pressure and varicocele development [10]. Although most cases occur on the left side, bilateral varicocele may develop when systemic or pelvic venous congestion is present [8]. Therefore, chronic venous congestion associated with Fontan circulation may be a risk factor for varicocele formation and deterioration of semen quality.

Meta-analyses have demonstrated that varicocelectomy in infertile men with clinically palpable varicocele significantly improves semen parameters compared with conservative

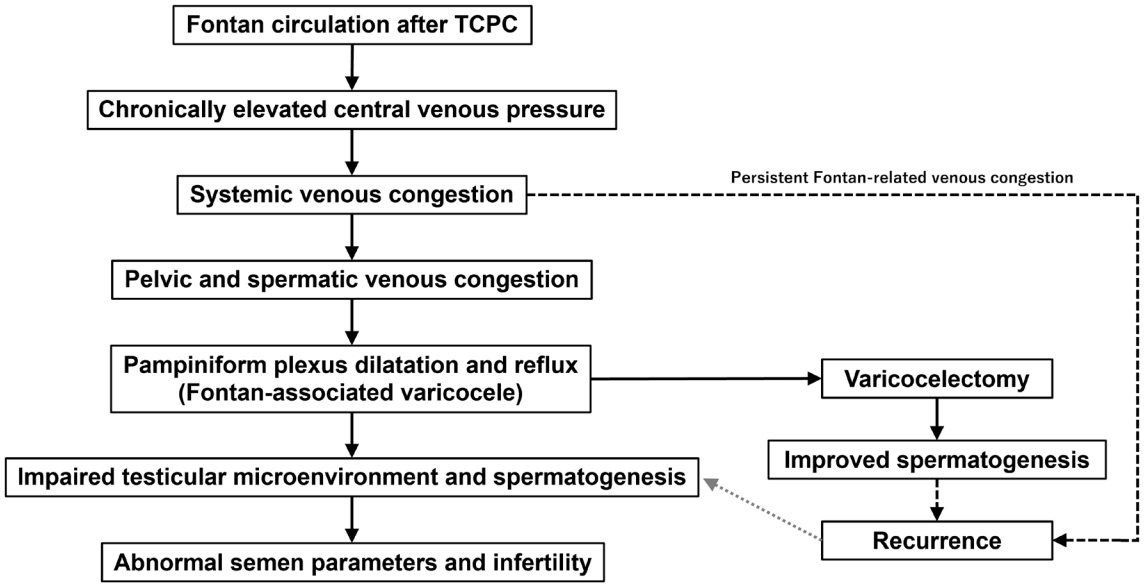

FIGURE 3 | Proposed mechanism of Fontan-associated varicocele, the potential effect of varicocelectomy, and possible recurrence.

management [11]. In Fontan patients, persistent venous hypertension may promote varicocele formation, impair spermatogenesis, and increase the risk of recurrence (Figure 3).

A recent meta-analysis also reported higher odds of sperm retrieval, sperm appearance in the ejaculate, and clinical pregnancy after varicocele repair before testicular sperm extraction [12]. In Case 1, varicocelectomy was selected because the couple did not wish to pursue ART. Semen parameters improved despite recurrent Grade 3 varicocele during follow-up, suggesting that morphological recurrence may not immediately restore the same adverse testicular environment. Even temporary improvement may be clinically meaningful in infertility treatment, particularly when timed with ART.

In these cases, perioperative management was discussed in advance with anesthesiology, cardiology, and cardiovascular surgery, because multidisciplinary coordination is essential in patients with Fontan circulation [13]. Particular attention should be given to the risks of thrombosis and bleeding in the perioperative period. Sperm DNA fragmentation was not evaluated because this test was not available at our institution at that time, which is a limitation of the present report. These cases also highlight the importance of careful preoperative evaluation in high-risk patients with Fontan physiology, and additional assessment of sperm quality may help in deciding the indication for surgery. Additional FAVC cases are needed to clarify its characteristics and its relationship to other conditions associated with elevated central venous pressure.

4 | Conclusion

We propose a new clinical entity, Fontan-associated varicocele, which develops as a consequence of the systemic venous congestion inherent to Fontan circulation. With careful perioperative management, varicocelectomy for FAVC may be effective in selected cases, although the risk of recurrence may be higher.

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During the preparation of this work the authors used ChatGPT in order to improve the language and clarity of the manuscript. After using this tool/service, the authors reviewed and edited the content as needed and take full responsibility for the content of the published article.

Ethics Statement

This study was approved by the institutional review board (approval number: K2504-003).

Consent

Written informed consent was obtained from both patients for publication of this case report and the accompanying images.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

References

1. P. Khairy, N. Poirier, and L. A. Mercier, "Univentricular Heart," *Circulation* 115, no. 6 (2007): 800–812, https://doi.org/10.1161/CIRCULATIONAHA.105.592378.

2. G. A. Mazza, E. Gribaudo, and G. Agnoletti, "The Pathophysiology and Complications of Fontan Circulation," *Acta Bio Medica: Ateneo Parmense* 92, no. 5 (2021): e2021260, https://doi.org/10.23750/abm.v92i5.10893.

3. B. W. Driesen, M. Voskuil, and H. B. Grotenhuis, "Current Treatment Options for the Failing Fontan Circulation," *Current Cardiology*

Reviews 18, no. 4 (2022): e060122200067, https://doi.org/10.2174/1573403X1866622010611451.

4. A. C. Egbe, Y. N. V. Reddy, A. R. Khan, et al., "Venous Congestion and Pulmonary Vascular Function in Fontan Circulation: Implications for Prognosis and Treatment," *International Journal of Cardiology* 271 (2018): 312–316, https://doi.org/10.1016/j.ijcard.2018.05.039.

5. C. de Lange, T. Möller, and H. Hebelka, "Fontan-Associated Liver Disease: Diagnosis, Surveillance, and Management," *Frontiers in Pediatrics* 11 (2023): 1100514, https://doi.org/10.3389/fped.2023.1100514.

6. A. B. Bhatt, M. J. Landzberg, M. Gerhard-Herman, E. Rodriguez-Huertas, D. Graham, and A. M. Valente, "Pathophysiology of Chronic Venous Insufficiency in Adults With a Fontan Circulation (a Pre-Defined Substudy of the CALF Investigation)," *International Journal of Cardiology* 165 (2013): 41–45, https://doi.org/10.1016/j.ijcard.2011.07.053.

7. K. N. Pundi, K. Pundi, J. N. Johnson, et al., "Contraception Practices and Pregnancy Outcome in Patients After Fontan Operation," *Congenital Heart Disease* 11, no. 1 (2016): 63–70, https://doi.org/10.1111/chd.12291.

8. C. L. Cho, S. C. Esteves, and A. Agarwal, "Novel Insights Into the Pathophysiology of Varicocele and Its Association With Reactive Oxygen Species and Sperm DNA Fragmentation," *Asian Journal of Andrology* 18, no. 2 (2016): 186–193, https://doi.org/10.4103/1008-682X.170441.

9. C. Munoz-Lopez, A. Wong, K. Lewis, R. Bole, S. C. Vij, and S. D. Lundy, "The Evolving Landscape of Male Varicocele Pathophysiology in the Era of Multi-Omics: A Narrative Review of the Current Literature," *Biology-Basel* 13, no. 2 (2024): 80, https://doi.org/10.3390/biology13020080.

10. H. L. Fred, "The Tricuspid Insufficiency-Pulsating Varicocele Connection: A Syndrome and Its History," *Texas Heart Institute Journal* 44, no. 5 (2017): 302–305, https://doi.org/10.14503/THIJ-17-6501.

11. A. Agarwal, R. Cannarella, R. Saleh, et al., "Impact of Varicocele Repair on Semen Parameters in Infertile Men: A Systematic Review and Meta-Analysis," *World Journal of Men's Health* 41, no. 2 (2023): 289–310, https://doi.org/10.5534/wjmh.220142.

12. S. Çayan, G. M. Pinggera, H. Alipour, et al., "The Effects of Varicocele Repair on Testicular Sperm Retrieval, Sperm Recovery in the Ejaculate and Clinical Pregnancy Rates in Non-Obstructive Azoospermic Men With Clinical Varicocele: A Systematic Review and Meta-Analysis," *World Journal of Men's Health* 43 (2025): e44, https://doi.org/10.5534/wjmh.250065.

13. J. R. McNamara, A. McMahon, and M. Griffin, "Perioperative Management of the Fontan Patient for Cardiac and Noncardiac Surgery," *Journal of Cardiothoracic and Vascular Anesthesia* 36, no. 1 (2022): 275–285, https://doi.org/10.1053/j.jvca.2021.04.021.