8th WORLD CONGRESS of PEDIATRIC SURGERY

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BILATERAL VIDEO-ASSISTED SINGLE-PORT TRANSUMBILICAL GONADECTOMY IN AN EXCEPTIONAL CONDITION: SWYER SYNDROME

Julio César Moreno Alfonso[1], Maria José Neira Morales[2], **Esther Comajuncosas Pérez**[1], Catalina Fonseca Rejtman[2], Isabella Garavis Montagut, Ada Molina Caballero[1], Alberto Pérez Martínez[1]

[1]Hospital Universitario de Navarra. Department of Pediatric Surgery. Pamplona; Spain
[2]Universidad El Bosque. Medical Faculty, Bogotá; Colombia

INTRODUCTION: Swyer syndrome, or pure **46 XY gonadal dysgenesis**, is a rare genetic condition associated with a high risk of gonadal neoplastic degeneration. Our experience with over 3,000 transumbilical video-assisted appendectomies has allowed us to safely extrapolate this technique to other indications. We describe a novel minimally invasive surgical approach for managing an exceptional gonadal pathology.

CASE PRESENTATION

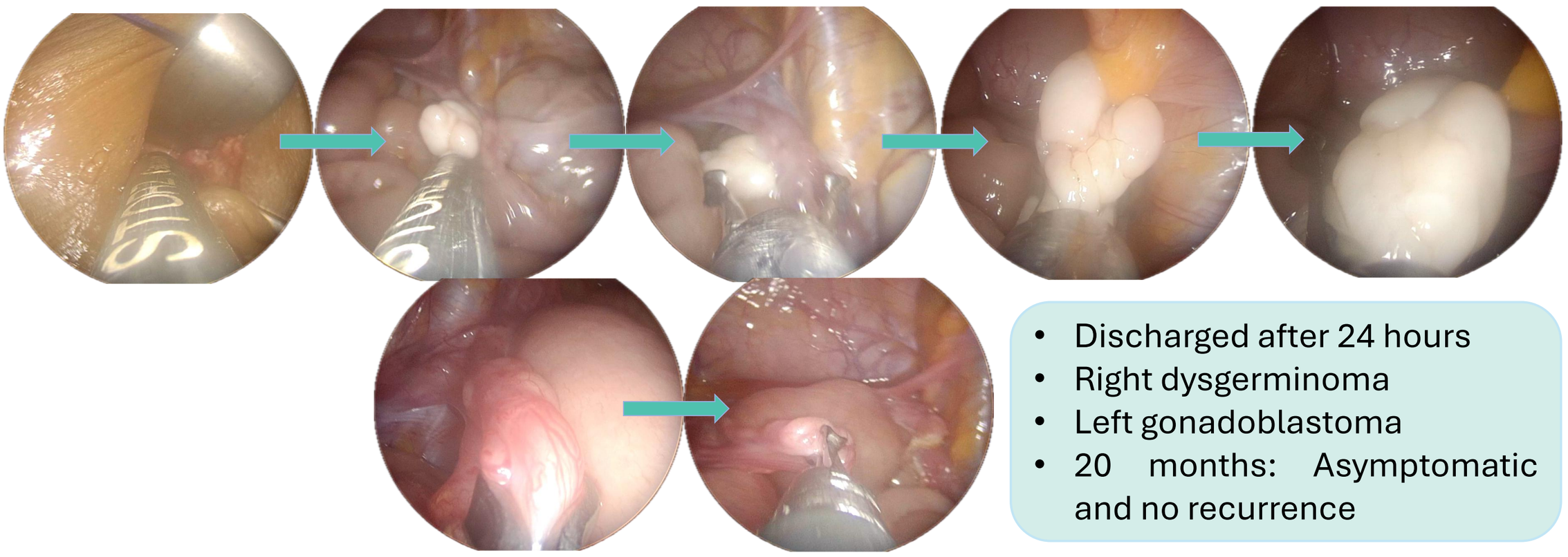
8 years old →Normal **female** phenotype → Karyotype 46 XY → Pure gonadal dysgenesis →Indication for **bilateral gonadectomy** → Risk of gonadal neoplasia

1. Umbilical incision and placement of surgical retractor →10 mm lens with working channel (5 mm)

2. Sealing of the pneumoperitoneum using a “tobacco pouch” suture

3. Identification, externalization, and gonadal excision through the navel

4. Peritoneal-muscle-aponeurotic closure in one piece →Imperceptible scar.



- Discharged after 24 hours
- Right dysgerminoma
- Left gonadoblastoma
- 20 months: Asymptomatic and no recurrence

CONCLUSION

Swyer syndrome is associated with a **high risk of gonadal malignancy**. This case is unique because it involves one of the youngest patients described in the literature and was treated using a **transumbilical video-assisted** approach, which combines the advantages of conventional **laparoscopy** and **open** surgery, with an excellent cosmetic result, leaving only a single scar on the navel.