

Herlyn Werner Wunderlich Syndrome with an Incidental Finding of Concomitant Ipsilateral Adrenal Agenesis

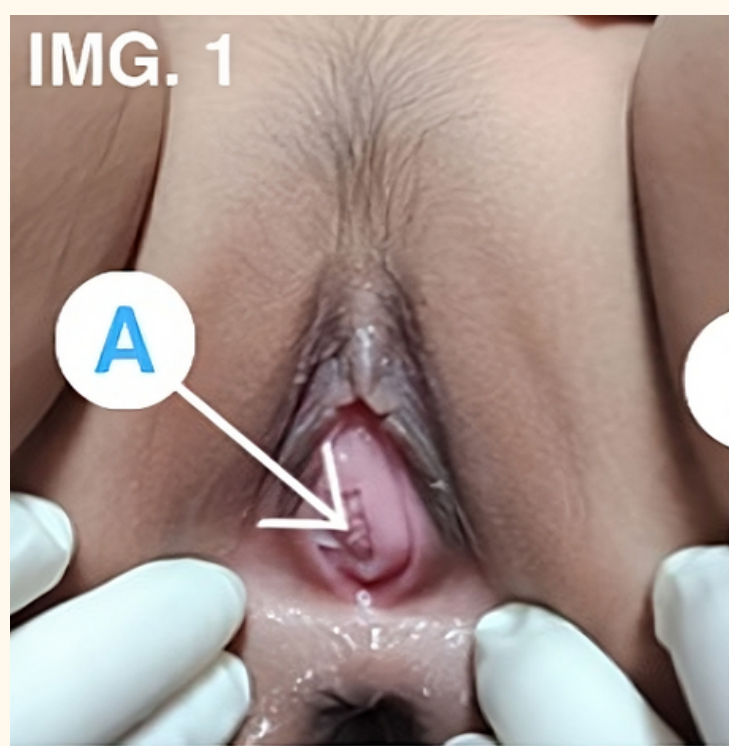
Rochelle Mae M. Supan, MD | Junette Grace A. Ko, MD, FPOGS, FPSGE

Department of Obstetrics and Gynecology
Davao Regional Medical Center, Apokon, Tagum City, Philippines

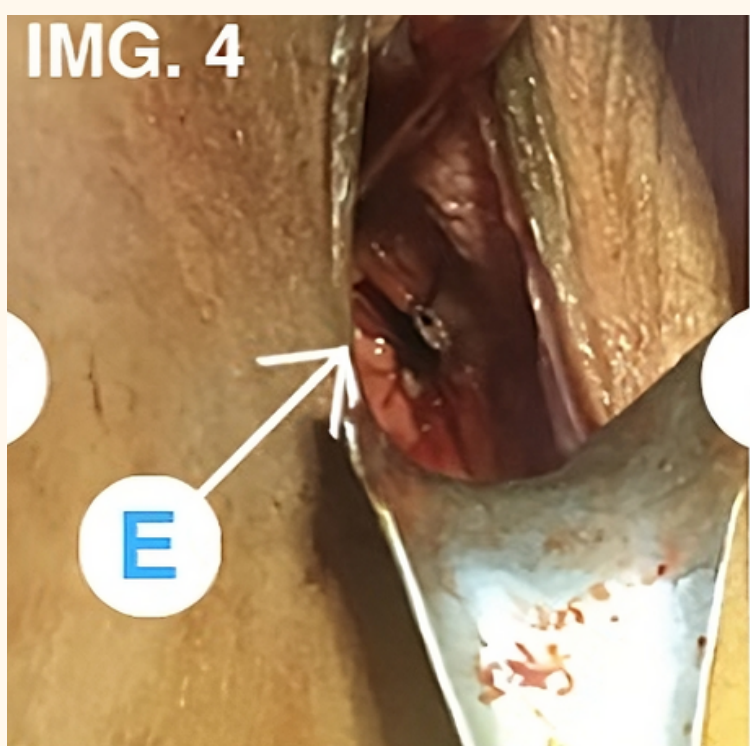
INTRODUCTION

Herlyn Werner Wunderlich Syndrome (HWWS) is a rare form of mullerian duct anomaly characterized by a combination of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. Internationally, its incidence is 0.1-3.8% and among the reported cases, none presented with a concomitant adrenal gland agenesis. This is the first case report of a diagnosed Herlyn Werner Wunderlich syndrome with a concomitant unilateral adrenal gland agenesis.

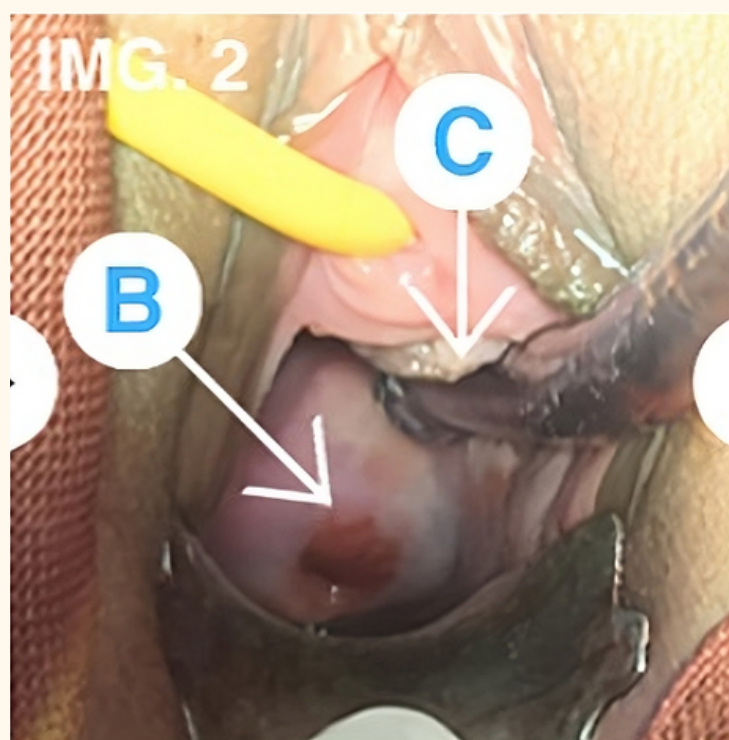
PROCEDURE



1 Patient's perineal examination showing a grossly normal vulva. The hymen is well estrogenized with a fleshy mass (Arrow A) bulging at the hymenal opening resembling that of the vaginal wall.



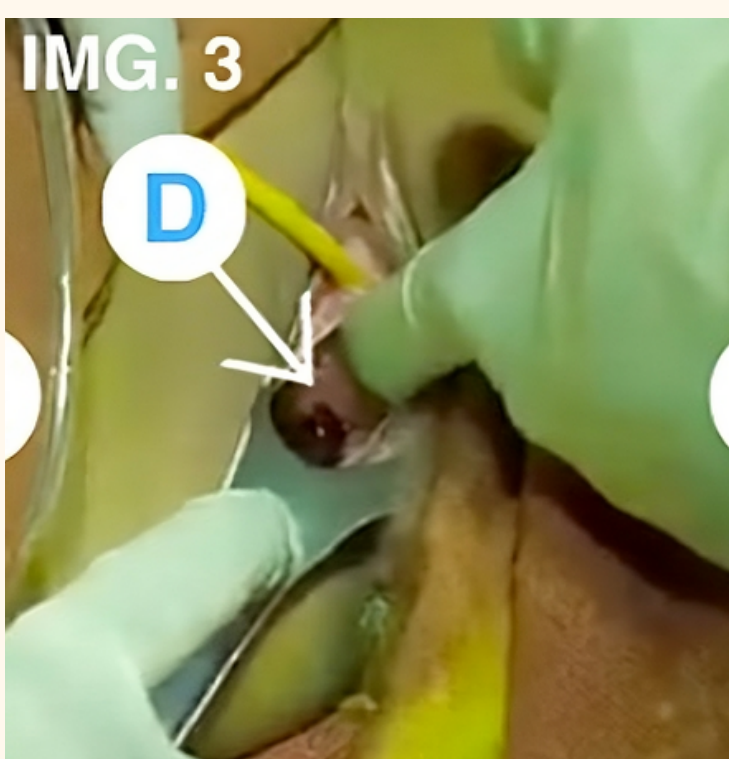
4 Post resection of the vaginal septum (Arrow E).



2 Inspection after regional anesthesia showing the unobstructed cervix (Arrow B) with the longitudinal vaginal septum displaced laterally (Arrow C).



5 Vaginal mold used daily for 2 weeks to prevent closure of the resected area.



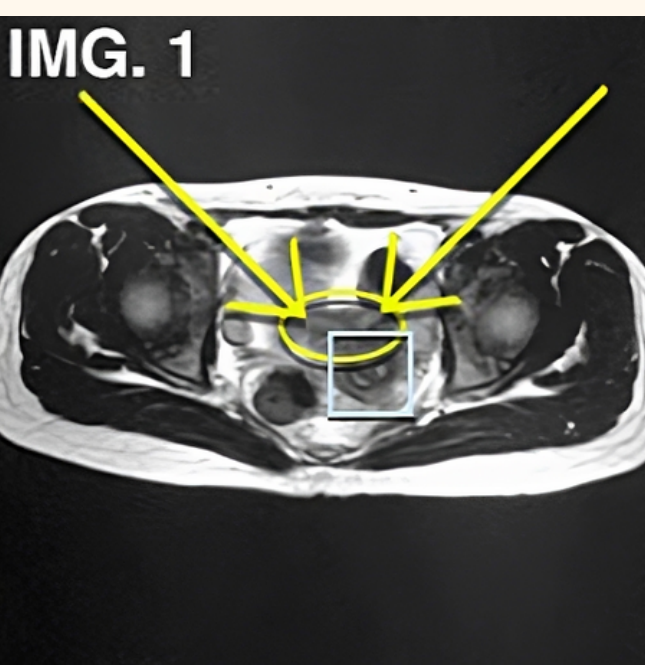
3 Resection of the vaginal septum was done by initially making an incision with the use of a stab knife subsequently followed by incising an approximately 2 x 2 cm of vaginal septum (Arrow D). After the resection, drainage of approximately 100 cc of hematocolpos was done.



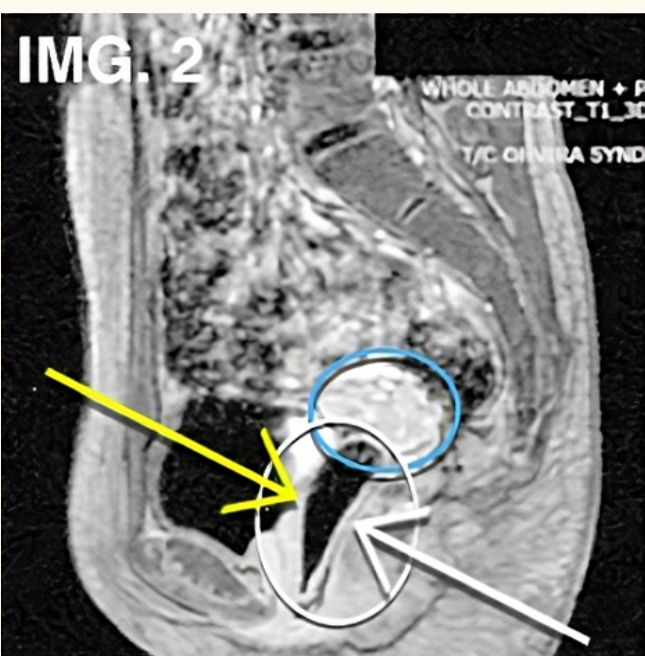
6 3D Transrectal Ultrasound, 6 months post resection of the longitudinal vaginal septum showing no recurrence of hematocolpos.

RESULTS

This is a case study of a 10-year old Filipino who presented with severe cyclic abdominal pain. MRI result showed two separate uteri and cervices, obstructed left hemivagina, absent left kidney, and adrenal gland. This patient has no symptoms of adrenal insufficiency, and serum cortisol and ACTH levels were within normal limits. She underwent vaginoscopy and resection of the longitudinal vaginal septum. Postoperatively, the regular menstrual cycle continued and was no longer associated with dysmenorrhea. Follow-up after six months revealed no recurrence of hematacolpos on 3D transrectal sonography.



Inside the yellow circle are the 2 separate uteri with divergent apices (pointed by yellow arrows). Inside the square are the 2 cervices.



Inside the white circle is the image of the obstructed left hemivagina (pointed by the yellow arrow on the 2nd image above) which is dilated and filled with fluid (blood). On the right side (pointed by the white arrow) is the collapsed unobstructed right hemivagina. The structure that is inside the blue circle is the uterus.

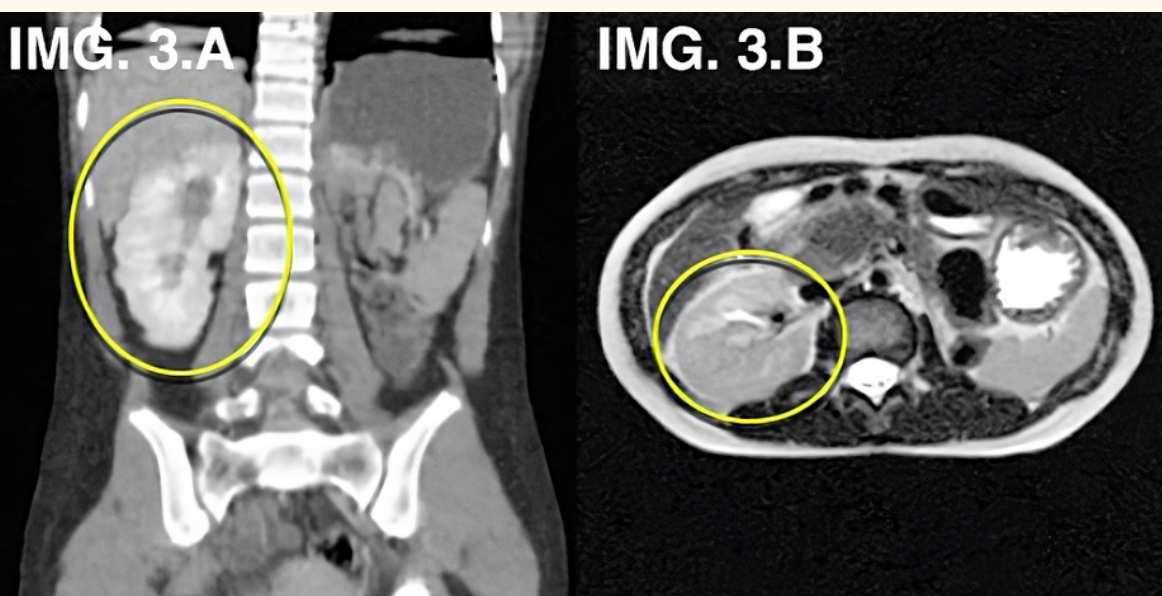
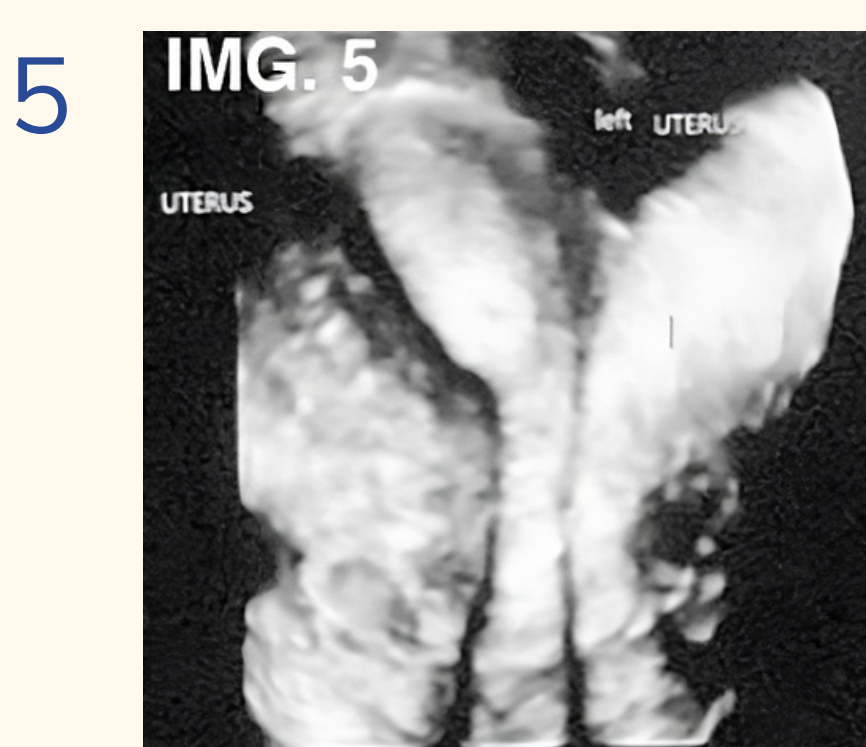


Image 3.A shows a coronal view of the abdominal MRI done on this patient, with note of an enlarged right kidney with normal parenchymal thickness and non-dilated ureter. The left kidney is absent. Image 3.B is an axial view of the MRI scan with right kidney (yellow circle) and absent left kidney.



This is an MRI slice that demonstrates an absent left adrenal gland. Inside the yellow circle is where the left suprarenal gland is supposed to be appreciated, if present.



3D Transrectal Ultrasound, 6 months post resection of the longitudinal vaginal septum showing no recurrence of hematocolpos.

CONCLUSIONS

The rarity of HWWS is a major contributory factor in delayed diagnosis; however, if detected early, timely decompression of vaginal obstruction could avoid complications and preserve reproductive capacity. A high index of suspicion is needed for patients who present with cyclic pelvic pain during puberty and patients who have findings of renal anomaly. The preferred surgical approach for these patients is resection of the vaginal septum since the main reason for the symptoms is the physical obstruction of the menstrual flow, which predisposes to endometriosis due to the retrograde flow of menstruation. This procedure is associated with a successful reproductive performance in the future. The significance of the incidental finding of concomitant adrenal agenesis in a diagnosed HWWS suggests that other abnormalities or defects may be associated with this syndrome and not just what is presently known. The congenital anomaly involving the reproductive organ poses a challenge in the patient's capacity to bring about a successful pregnancy. Adding to that is the possibility of developing adrenal insufficiency during stressful situations or pregnancy. It is crucial to consider these things in managing this patient. The body of knowledge is ever-changing and fast-growing. What we know now may only be a fraction of what will be discovered in the future. This case report hopes to be a beacon which will pave a way for future studies that is more focused on unilateral gland agenesis.