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Dienogest use for recurrent pyosalpinx as a long-term complication of cloacal exstrophy: A case report

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Abstract

Background

Cloacal exstrophy (CE) is a rare congenital disease that requires multiple surgeries for complex gastrointestinal and genitourinary anomalies. Long-term complications are not uncommon; however, they are poorly reported. Pyosalpinx is sometimes encountered during CE management in adolescents and young adults.

Case

A 28-year-old woman with a history of CE presented with fever, lower abdominal pain, and vomiting and was diagnosed with left pyosalpinx. Computed tomography-guided drainage and intravenous antibiotic administration were successful; however, she had 2 readmissions for recurrent pyosalpinx 1 week after discharge and again 4 months later. She was administered Dienogest, a synthetic progestin, to prevent recurrent pyosalpinx and had no recurrence for 8 months.

Summary and conclusion

Dienogest is a conservative treatment choice for preventing the recurrence of pyosalpinx for CE patients.

Key Words: Cloacal exstrophy; Dienogest; Long-term complication; Pelvic

19 inflammatory disease; Pyosalpinx

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Introduction

Cloacal exstrophy (CE) is a rare congenital condition observed in 1 of 200,000-400,000 live births (1, 2). CE remains a challenging disease compromising multiple gastrointestinal, genitourinary, and skeletal anomalies; however, its survival rate is now approximately 100% resulting from improved neonatal care and surgical management (1).

Conversely, CE patients experience many genitourinary and gastrointestinal problems from childhood to young adulthood. In particular, most females with CE are prone to gynecological complications throughout life due to congenital anomalies of the vagina and uterus, and their long-term outcomes are poorly described in living adults (2, 3).

Pyosalpinx is a severe sequela of pelvic inflammatory disease (PID) in which the fallopian tubes are filled with pus. PID is an ascending infection from the vagina or cervix to the upper genital tract and is attributed to *Chlamydia trachomatis*, *Neisseria gonorrhoeae*, bacterial vaginosis, and enteric pathogens (4). CE patients have been reported to experience recurrent PID, such as pyosalpinx or tubo-ovarian abscess (TOA) and are often difficult to treat (3, 5).

Here, we present a case of a young woman with recurrent pyosalpinx following CE management in childhood and adolescence. Dienogest (DNG), a synthetic progestin, was effective in controlling recurrent pyosalpinx; thus, the pathogenesis of recurrent

pyosalpinx in CE patients and the efficacy of DNG are discussed.

Case

A 28-year-old woman with a history of CE presented with fever, lower abdominal pain, and vomiting and was transferred to our hospital with a diagnosis of left pyosalpinx. She was born with a weight of 2622 g at 36 weeks of gestation by cesarean section and was diagnosed with CE. She underwent primary closure of CE and ileostomy on day 4 of life and continent urinary diversion by right to left transureteroureterostomy with cutaneous right ureterostomy, augmentation cystoplasty using an ileal patch, and intestinal vaginoplasty at the age of 5 years. Her first menstruation was at age 12, but due to dysmenorrhea, she underwent a right hemi-hysterectomy for uterine didelphys and left uterine to fallopian tube anastomosis at the age of 15 years. After that, her condition was well controlled in the outpatient clinic without medication. She managed her bowel movements with colostomy and required clean intermittent catheterization four times a day. She was sexually active and had sexual intercourse with her partner one week before admission.

Laboratory evaluation at admission revealed a C-reactive protein level of 17.52 mg/dl and a creatinine level of 1.07 mg/dl, suggesting acute kidney injury. Enhanced abdominal

computed tomography (CT) showed a 79×52 mm left pyosalpinx and multiple small abdominal abscesses, leading to bilateral hydronephrosis and vomiting due to intestinal obstruction (**Fig. 1**). CT-guided drainage was performed for the left pyosalpinx. The brown pus contained *Streptococcus anginosus*, *Streptococcus agalactiae*, and *Bacteroides fragilis*. Samples collected from the cervix tested negative for *Chlamydia trachomatis* and *Neisseria gonorrhoeae*. Renal function improved within normal limits, and symptoms of fever, abdominal pain, and vomiting disappeared a few days after drainage. Conservative antibiotics (intravenous tazobactam/piperacillin and penicillin G for a total of 2 weeks; thereafter, oral amoxicillin/clavulanate) were administered successfully, the drain was removed on day 21, and she was discharged on day 25.

She had 2 readmissions which were 1 week after discharge, and then again 4 months later for recurrent pyosalpinx. During these hospitalizations, only intravenous antibiotics were administered. Abdominal ultrasonography (US) at the second hospitalization showed hydrosalpinx and anastomotic stenosis between the left uterus and left fallopian tube, which suggested a cause of recurrent pyosalpinx (**Fig. 2A**). A transvaginal approach using hysteroscopy for releasing the anastomotic stenosis between the left uterus and fallopian tube was considered; however, it was difficult due to a complicated surgical history. Therefore, the decrease in menstrual fluid in the left fallopian tube may have prevented

recurrent pyosalpinx, and in the third hospitalization, she started DNG at 2 mg daily orally. After DNG administration, fluid collection in the left fallopian tube completely disappeared on abdominal US (**Fig. 2B**), and she had no recurrence for 8 months after DNG administration.

Summary and conclusion

In this case, a young woman following CE management in childhood and adolescence suffered from recurrent pyosalpinx, and DNG, a synthetic progestin, was effective in preventing recurrent pyosalpinx. Only one young adolescent girl with a history of CE has been reported to experience pyosalpinx and TOA and undergo a radical hysterectomy (5). Thus, this is the first report to treat pyosalpinx as a long-term complication during CE management successfully.

CE is a challenging disease that requires multiple surgeries for complex gastrointestinal, genitourinary, and skeletal anomalies. Because of the improvements in neonatal care and surgical techniques, the survival rate has been approaching 100% in the last few decades (2, 3). However, as long-term survivors of CE have become more common, CE patients have been found to suffer from many gynecological disorders. A recent systematic review reported that 57.1% of female CE patients had vaginal-related issues and that 14.3% to

71.0% had uterine anomalies, such as uterine didelphys and uterine bipartitus (2). Owing to these anomalies, many females with CE experienced dysmenorrhea during puberty, with related gynecologic surgery performed in approximately two-thirds of cases (2). In our case, right hemi-hysterectomy and left uterus-fallopian tube anastomosis were performed for the management of dysmenorrhea, and recurrent pyosalpinx occurred as a long-term complication of the anastomosis.

On the other hand, regarding upper genital tract tissues, including PID such as pyosalpinx and TOA in CE patients, no detailed observational studies and only limited case reports have been found (5). PID is one of the most common gynecological problems in young women (4), and it must be considered a major problem that reduces quality of life, especially in CE patients.

Pyosalpinx is a severe form of PID in which the fallopian tube is filled with pus. Treatment of pyosalpinx ranges from conservative intravenous antibiotics and image-guided drainage to laparoscopic aspiration, salpingostomy, and salpingectomy.

Urogenital anomalies, including CE, have been reported to be one of the risk factors for pyosalpinx, and surgical treatment is performed in most cases (5, 6). In our case, the primary treatment for pyosalpinx was successful by CT-guided drainage and intravenous antibiotics administration; however, pyosalpinx recurred twice. On the abdominal US

findings at recurrence, the cause of recurrent pyosalpinx was suspected to be left hydrosalpinx resulting from stenosis of the uterus-fallopian tube anastomosis, which is a long-term complication of CE management.

DNG is a unique fourth-generation synthetic progestogen mainly used for the long-term management of endometriosis worldwide. It is considered effective in decreasing the size of endometriomas and reducing endometriosis-associated pain (7). In addition, it has less severe side effects, such as abnormal uterine bleeding and headache, with long-term use compared with other progestin products (8). In cloacal anomalies, ovarian function is normal; thus, ovary-releasing estrogen stimulates the endometrium in the uterus after puberty, and some experience menstrual flow obstruction (3). Hormonal suppression of endometrial stimulation and menses prevents the continued accumulation of obstructed menstrual products in CE patients (3). In this patient, hormonal suppression by DNG may have played an important role in reducing menstrual blood flow, which contributed to reduced fluid accumulation in the left fallopian tube and controlled recurrent pyosalpinx without severe side effects.

In conclusion, patients with CE often experience gynecological problems as long-term complications. Pyosalpinx in patients with CE is a major issue that can lead to multiple hospitalizations and reduce the patient's quality of life. DNG is a conservative treatment

129 option for recurrent pyosalpinx as a long-term complication of CE.

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Figure 1 Enhanced abdominal CT findings at diagnosis

A: Axial imaging showing a 79×52 mm left pyosalpinx on the back of the bladder and multiple small abdominal abscesses.

B: The coronal imaging revealed bilateral hydronephrosis associated with compression by the pyosalpinx.

CT, computed tomography.

Figure 2 Abdominal US findings before and after DNG administration

A: Before DNG administration, abdominal US showed left hydrosalpinx and suspected anastomotic stenosis between the left uterus and left fallopian tube.

B: Under DNG administration, no fluid collection in the left fallopian tube was observed on abdominal US.

DNG, Dienogest; US, ultrasonography.



