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

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Patient consent is not required because no personal information or details are included.

1 **Abstract**

2 **Background**

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

7 **Case**

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

14 **Summary and conclusion**

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

17

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

19 inflammatory disease; Pyosalpinx

20

21 **Introduction**

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

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

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

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

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

40

41 **Case**

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

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

57 computed tomography (CT) showed a 79×52 mm left pyosalpinx and multiple small  
58 abdominal abscesses, leading to bilateral hydronephrosis and vomiting due to intestinal  
59 obstruction (**Fig. 1**). CT-guided drainage was performed for the left pyosalpinx. The  
60 brown pus contained *Streptococcus anginosus*, *Streptococcus agalactiae*, and  
61 *Bacteroides fragilis*. Samples collected from the cervix tested negative for *Chlamydia*  
62 *trachomatis* and *Neisseria gonorrhoeae*. Renal function improved within normal limits,  
63 and symptoms of fever, abdominal pain, and vomiting disappeared a few days after  
64 drainage. Conservative antibiotics (intravenous tazobactam/piperacillin and penicillin G  
65 for a total of 2 weeks; thereafter, oral amoxicillin/clavulanate) were administered  
66 successfully, the drain was removed on day 21, and she was discharged on day 25.  
67 She had 2 readmissions which were 1 week after discharge, and then again 4 months later  
68 for recurrent pyosalpinx. During these hospitalizations, only intravenous antibiotics were  
69 administered. Abdominal ultrasonography (US) at the second hospitalization showed  
70 hydrosalpinx and anastomotic stenosis between the left uterus and left fallopian tube,  
71 which suggested a cause of recurrent pyosalpinx (**Fig. 2A**). A transvaginal approach using  
72 hysteroscopy for releasing the anastomotic stenosis between the left uterus and fallopian  
73 tube was considered; however, it was difficult due to a complicated surgical history.  
74 Therefore, the decrease in menstrual fluid in the left fallopian tube may have prevented



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

79

## 80 **Summary and conclusion**

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

87 CE is a challenging disease that requires multiple surgeries for complex gastrointestinal,  
88 genitourinary, and skeletal anomalies. Because of the improvements in neonatal care and  
89 surgical techniques, the survival rate has been approaching 100% in the last few decades  
90 (2, 3). However, as long-term survivors of CE have become more common, CE patients  
91 have been found to suffer from many gynecological disorders. A recent systematic review  
92 reported that 57.1% of female CE patients had vaginal-related issues and that 14.3% to

93 71.0% had uterine anomalies, such as uterine didelphys and uterine bipartitus (2). Owing  
94 to these anomalies, many females with CE experienced dysmenorrhea during puberty,  
95 with related gynecologic surgery performed in approximately two-thirds of cases (2). In  
96 our case, right hemi-hysterectomy and left uterus-fallopian tube anastomosis were  
97 performed for the management of dysmenorrhea, and recurrent pyosalpinx occurred as a  
98 long-term complication of the anastomosis.

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

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

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

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

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

126 In conclusion, patients with CE often experience gynecological problems as long-term  
127 complications. Pyosalpinx in patients with CE is a major issue that can lead to multiple  
128 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.

130

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153 **Figure 1 Enhanced abdominal CT findings at diagnosis**

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

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

158 CT, computed tomography.

159

160 **Figure 2 Abdominal US findings before and after DNG administration**

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

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

165 DNG, Dienogest; US, ultrasonography.





