

Persistent Cloaca: Lessons Learnt from a Case

V Shakya, S Priyadarshini, R Koirala, S Khaniya

Citation

V Shakya, S Priyadarshini, R Koirala, S Khaniya. *Persistent Cloaca: Lessons Learnt from a Case*. The Internet Journal of Surgery. 2008 Volume 18 Number 1.

Abstract

Persistent cloaca is a rare congenital anomaly. It has seldom been reported from our country. Due to the complexity of its anatomy, surgeons have found it difficult to handle. There are many controversies regarding its management. With a case report, we try to discuss briefly the fallacies in management and current recommendations in the initial newborn period.

INTRODUCTION

A persistent cloaca is a complex anorectal and genitourinary malformation, in which the rectum, vagina, and urinary tract meet and fuse, creating a single common channel. Cloacas occur in 1 of every 40,000-50,000 live births. Cloacas appear in a wide spectrum of variation. The single orifice, called a common channel, may occur varying in length from 1 to 10cm. However, in the newborn period, there is still a lot of controversy regarding the initial management. We had one case of persistent cloaca that was managed in the neonatal period; however, we could not save her. With this case report we try to discuss various options in its management and potential mishaps.

CASE REPORT

A 2-day-old female child was brought to our hospital with failure to pass meconium since birth and absence of anal opening. On examination, she was found to have a distended abdomen and a single perineal opening (Fig. 1). She was subjected to an abdominal radiograph which showed a mass in the lower abdomen displacing the intestines to the periphery (Fig. 2). An ultrasound of the abdomen was also done which showed a distended bladder, a distended vagina, the loop of sigmoid and bilateral hydronephrosis (Fig. 3). Finally, at laparotomy, we found a dilated urinary bladder, an over-distended uterus, and a dilated sigmoid colon, which all merged into the pelvis (Fig. 4). A decompressing divided colostomy at the lower part of the descending colon along with a suprapubic cystostomy was done. Both functioned well; however, the child continued to be lethargic, had high fever and had a persistent lump in the lower abdomen. Re-ultrasound suggested it to be the distended uterus, and she had persistent hydronephrosis. The child died on the 3rd

postoperative day.

Figure 1

Figure 1: Perineum of the child showing a single opening



Figure 2

Figure 2: A supine abdominal radiograph shows hydrocolpos (arrow) displacing the intestines to the periphery

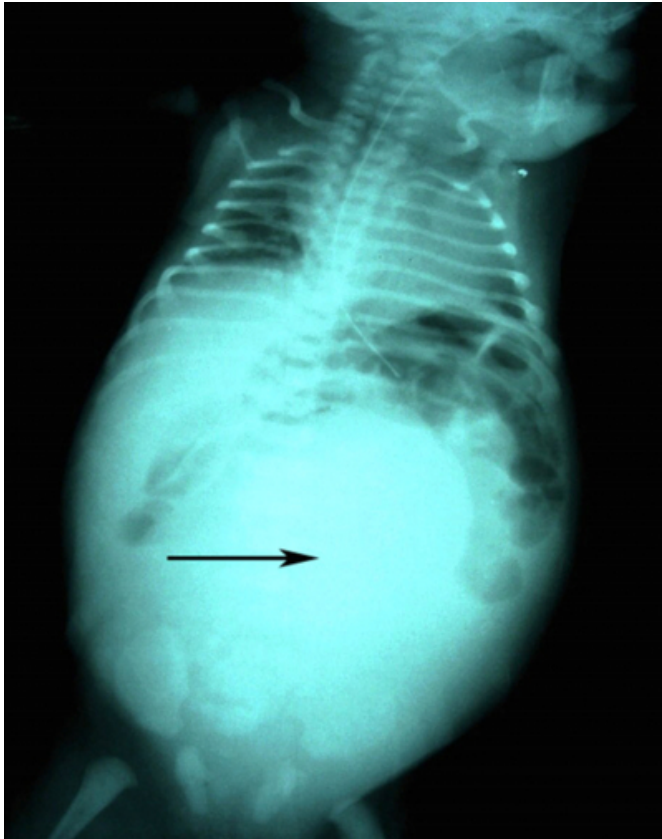


Figure 3

Figure 3: An abdominal ultrasound in figure 3a showing the dilated bladder (white arrow), the distended uterus (white arrowhead) and the loop of sigmoid colon (black arrows); and in figure 3b showing the hydronephrotic kidneys (white arrows) on both sides of the spine.

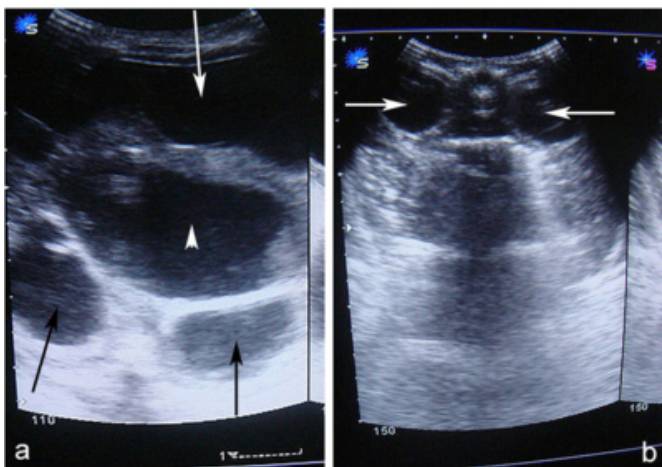
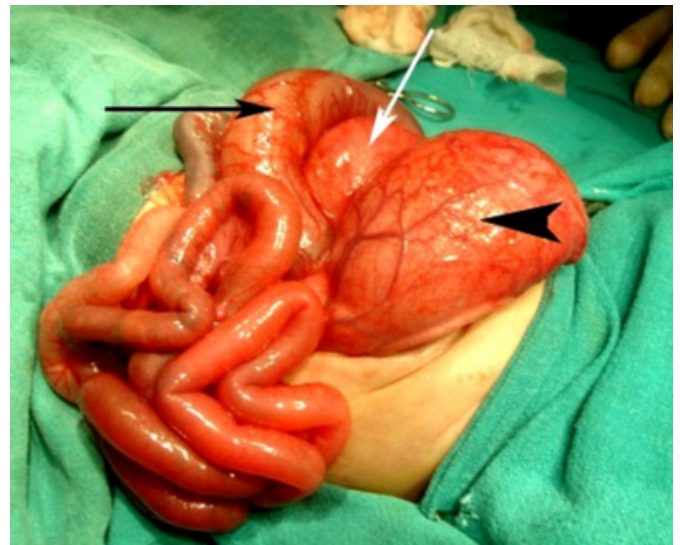


Figure 4

Figure 4: Laparotomy showing the sigmoid colon (black arrow), the distended uterus (white arrow) and the dilated bladder (arrowhead)



DISCUSSION

Cloacal anomalies are complex problems and rare anomalies. They are the most challenging of anorectal malformations and make up 13.6% of this group. ¹ Definitive repair has been reported earlier from our region; however, difficulties persist even earlier in the newborn period before these children get a chance for definitive repair. ²

Examination of a child with persistent cloaca begins with examination of the abdomen. On abdominal examination, a suprapubic mass may be present due to distended bladder or hydrometrocolpos or both. The abdominal distension may be severe due to hydrometrocolpos, bladder and/or intestinal distension. The next step is inspection of the perineum, which shows a single perineal opening. Inadequate perineal examination has led to false diagnosis of rectovestibular or rectovaginal fistula, leading thereby to a repair with mobilization of the rectum, only leaving the urogenital sinus untouched. ³ Only a colostomy may be done, and this can have fatal implications which lead to obstructive uropathy, sepsis, acidosis, and, sometimes, death.

Radiological evaluation begins with an abdominal radiograph, not an invertogram. The radiograph shows a ground glass appearance in the lower abdomen signifying a large abdominal mass which can be hydrocolpos or a distended bladder, displacing the intestines to the periphery. Next is ultrasonography of abdomen and pelvis which shows hydroureteronephrosis, hydrocolpos and the distended urinary bladder.

As a part of management, in our case, a divided colostomy at the lower part of the descending colon and a suprapubic cystostomy was done. Though both were functioning properly, the child had a persistent lump in the suprapubic region and hydroureteronephrosis. The vagina must have failed to drain properly; the hydrocolpos resulted probably into bilateral hydronephrosis, urosepsis and death.

The goal of the early management in the neonatal period is to detect associated anomalies, divert the gastrointestinal tract and divert the urinary tract properly. Fecal diversion can be in the form of loop colostomy, but nowadays, a colostomy placed in the descending colon and with separated stomas has been recommended.^{4,5} The stomas should be separated enough so that there is no spillage of the fecal contents to the distal loop, which minimizes the risk of urinary tract infection and sepsis. There also needs to be adequate distal bowel for the future pull-through. Authors now recommend placing the colostomy in the descending colon just after the colon takes off from its left retroperitoneal attachments, which is a relatively fixed part of the colon.^{5,6} This has been found to significantly reduce the incidence of prolapse. Prolapse leading to resection of bowel is potentially dangerous in these patients because it decreases the length of bowel available for pullthrough and might loosen the consistency of the stool increasing the chance of fecal incontinence.

Another anomaly to tackle is the genitourinary system. The bladder is frequently distended, and vesicostomy/suprapubic cystostomy is done to decompress the bladder. However, the distended vagina also creates pressure on the trigone

resulting in hydronephrosis.^{7,8} So it is recommended that a tube vaginostomy should also be done to decompress them and thus prevent complications, such as pyocolpos or ureteral obstruction.⁵ It has also been suggested that if the vagina is large enough to reach the umbilicus, it can be sutured to the abdominal skin like a colostomy.⁵

With this report and a review, we have tried to highlight common mistakes that are apt to happen in the neonatal period and current recommendations. Before definitive repair, these children have to be managed properly in the neonatal period according to recent guidelines.

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Author Information

Vikal Chandra Shakya, M.S. (Gen. Surg.)

Senior Resident, Department of Surgery, B. P. Koirala Institute of Health Sciences

Sugandha Priyadarshini, M.S. (Radiology)

Senior Resident, Department of Radiology, B. P. Koirala Institute of Health Sciences

Rabin Koirala, M.S. (Gen. Surg.)

Assistant Professor, Department of Surgery, B. P. Koirala Institute of Health Sciences

Sudeep Khaniya, M.S. (Gen. Surg.)

Assistant Professor, Department of Surgery, B. P. Koirala Institute of Health Sciences