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CASE REPORT

# Adenocarcinoma in isolated rectal bladder after treatment of bladder exstrophy

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**Abstract**
We report the first Finnish patient with carcinoma in an augmented intestinal bladder, where urine and stools are not in contact. The patient had undergone rectal bladder reconstruction at the age of 2 years because of bladder exstrophy. When the patient was aged 46 years, a 2-cm, papillar, well-differentiated adenocarcinoma was detected and removed, preserving the rectal bladder.

**Key Words:** *Carcinoma, bladder exstrophy, bladder augmentation, bladder replacement, enterocystoplasty, rectal bladder*

## Introduction

Stones, metabolic acidosis, infections, perforations and carcinoma are well-recognized complications of an augmented intestinal bladder [1]. However, the risk of carcinoma is not yet known and we feel it is of utmost importance to report all cases of carcinoma in intestinal bladders. Herein we report the first patient in Finland to develop carcinoma in an intestinal bladder, in which urine and stools are not in contact.

## Case report

At the age of 2 years, the bladder of a male patient was removed and the rectal bladder reconstructed because of bladder exstrophy. During the operation the ureters were connected to the rectum. The rectum itself was divided and was left as a urinary bladder. The distal end of the sigma was brought to the ventral side of the anus through the common sphincter mechanism [2]. At the end of the operation the patient had two anal orifices: the ventral one for stools and the dorsal one for urine. Stools and urine were not in contact. The patient experienced occasional bacteriuria, but apparently he did not have symptomatic urinary tract infections during childhood. The patient was satisfied with his urinary continence and with the ability to void spontaneously. Because of the anal stricture he needed occasional enemas in order to defecate.

At the age of 42 years the patient started to experience recurrent pyelonephritis with left flank pain. Scintigraphy revealed that the function distribution was 70% on the right side and 30% on the left. An obstruction was observed in the left rectoureteral junction and was treated with antegrade balloon dilatation. No endoscopy of the rectal bladder was performed on that occasion.

At the age of 46 years the patient was admitted from another part of the country to the Hospital for Children and Adolescents, Helsinki, where the original operations were performed, to discuss the primary diagnosis and the childhood operations. It appeared that the patient had undergone no endoscopic follow-up of the rectal bladder. During the same visit, endoscopy of the rectal bladder revealed a 2-cm papillary tumor. At biopsy a well-differentiated adenocarcinoma was detected, but there was no evidence of tumor invasion or metastases (Figure 1). The tumor was removed in the adult urology unit with rectum-preserving electroresection.

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During a 3-year follow-up period, in addition to a couple of CT scans, the patient underwent endoscopy of the rectal bladder, urine cytology and plasma carcinoembryonic antigen controls every 4 months during the first year and every 6 months thereafter. At the last follow-up session in June 2007 there was no evidence of tumor recurrence, no incontinence and the patient was still able to void spontaneously.

## Discussion

The reported risk of carcinoma after ureterosigmoidostomy (in which stools and urine mix) varies between 6% and 29% and the average interval between ureterocolostomy and the development of tumor is 20–26 years [3]. Carcinoma may result from the nitrosoamides produced by bacterial action in the mixture of urine and stools [4] or from active oxygen radicals that are produced by phagocytic cells as a result of inflammation at the anastomotic site [5].

During the last 30 years, intestinal bladder augmentation has largely replaced ureterosigmoidostomy. One reason for this has been to prevent urine and stools from mixing in the hope of reducing the risk of carcinoma. However, carcinomas have also been reported after intestinal bladder augmentation. The suggested risk of carcinoma in an augmented bladder is 3.6% after 25 years [6]. To date >30 cases of carcinoma have been published, consisting mostly of adenocarcinomas or transitional cell carcinomas [6]. Because of the long latency period for the development of cancer the risk remains uncertain. Stools and urine were not mixing in our patient and the situation was very similar to that after bladder augmentation or replacement nowadays.

In 1944 the incidence of bladder carcinoma in bladder exstrophy was estimated to be 4% [7]. It seems that successful operations to close the exstrophied bladder at an early age prevent carcinoma. In recent decades, bladder carcinoma in bladder exstrophy has become rare. Bladder reconstruction, however, is not always successful enough to prevent incontinence and 0–70% of children need bladder augmentation or replacement [8].

In our case, the patient was satisfied with his urinary continence and with the ability to void spontaneously. He was against complete resection of the rectal bladder and did not like the idea of a new intestinal bladder with a continent stoma. Because there was no sign of invasion and the adenocarcinoma was well differentiated, organ-sparing surgery was considered sufficient [9]. During a 3-year follow-up period there has been no sign of tumor recurrence.

Reported tumor cases have been rare after intestinal bladder augmentation or replacement, but their incidence is probably increasing. Currently, direct visualization by means of endoscopy is the only reliable method for finding carcinoma inside the intestinal bladder. A lifetime annual surveillance program, including cystoscopy and cytology, is recommended, starting 8–10 years after the augmentation [3,10]. Currently, in our institution we start cystoscopy controls earlier, so as to also monitor benign conditions such as stones.

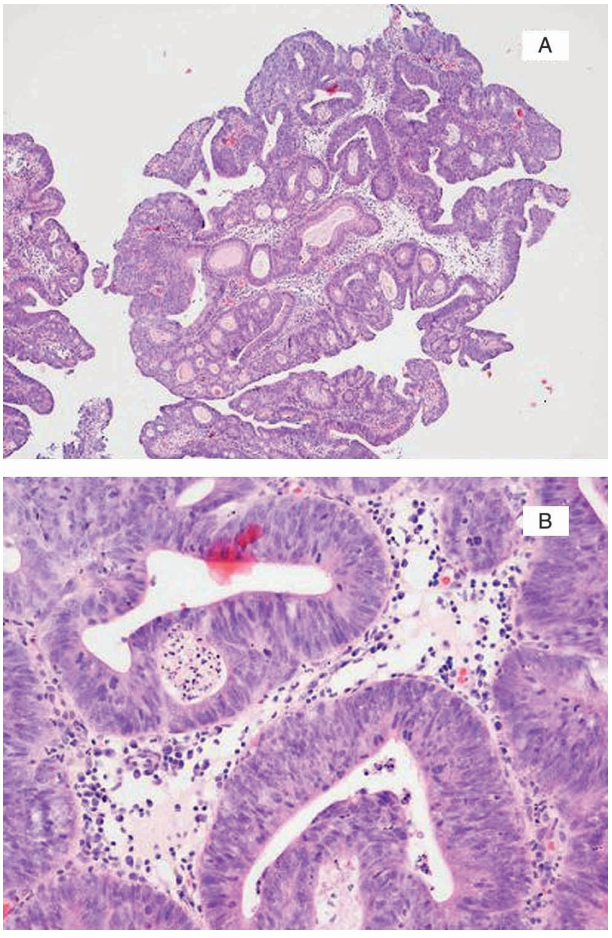


Figure 1. (A) Low-magnification image showing the polypoid architecture of the tumor. Histology is that of an adenocarcinoma of intestinal type. (B) High-magnification image showing marked atypia of the neoplastic epithelial cells and numerous mitoses.

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