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External and internal-external biliary drainage in children with malignant obstructive jaundice

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Abstract *Background.* Obstructive jaundice is an uncommon but important clinical problem in children with cancer. Percutaneous transhepatic biliary drainage is widely used to relieve malignant biliary obstruction in adults, but its use in children has not been well described.
Materials and methods. Six patients aged between 1 and 17 years underwent external or internal-external biliary drainage to relieve malignant obstructive jaundice. Biochemical, haematological and microbiological parameters were measured before the procedure and repeated 7–9 days later.
Results. External or internal-external biliary drainage was technically successful in all patients. No patient developed clinically significant biliary sepsis. Asymptomatic duodenal perforation occurred in one patient with bulky duodenal tumour following conversion from external to internal-external drainage.
Conclusions. External biliary drainage may be successfully performed in children with malignant obstructive jaundice.

Introduction

Malignant obstructive jaundice in children and adolescents presents quite different clinical problems to those encountered in adults [1]. In particular, most paediatric tumours will show significant reduction in size in response to therapy. Most malignant obstructive jaundice in children is, therefore, likely to be temporary. External biliary drainage (EBD) or internal-external biliary drainage (IEBD) may achieve relief of biliary obstruction until chemotherapy or other treatment takes effect.

Materials and methods

This paper reports the results of EBD and IEBD in all patients treated for malignant bile duct obstruction at two paediatric cancer centres between 1995 and 1998. The medical charts and radiological records were reviewed. Two patients were recruited retrospectively; the other four were treated after the study commenced.

Any coagulopathy was corrected before EBD and broad-spectrum antibiotic cover was used in all patients. EBD was performed following percutaneous transhepatic cholangiography (PTC). Access to the biliary tree was obtained by needle puncture of a bile duct, using sonographic or fluoroscopic guidance (Table 1, Fig. 1a). In one patient, a previously inserted nasobiliary drain permitted cholangiographic guidance. A 6-F or 8-F pigtail catheter was inserted over a guidewire into the bile ducts and positioned so that the distal loop of the catheter was formed above the level of obstruction (Fig. 2a). In one patient, it was technically easy to cross the obstruction with a guidewire and catheter and IEBD was performed as a first procedure (Fig. 1b).

In three patients, the drainage catheter was subsequently replaced by one or two IEBD catheters (Table 1, Figs. 2b–d). In one patient, EBD was converted to a permanent stent. Transbiliary forceps biopsy was performed in one patient at the same procedure as exchange of IEBD catheters.

Biochemical, haematological and microbiological parameters measured before and 7–9 days after the procedure were compared to assess the success and safety of the procedure. Subsequent clinical follow-up was performed to identify any complications.

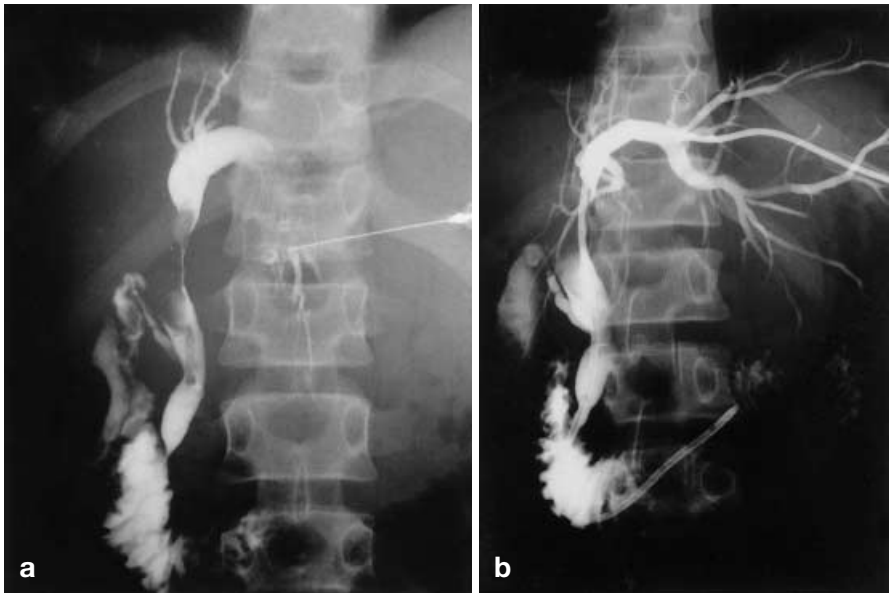

Fig. 1 a, b A 16-year-old male with hepatocellular carcinoma (case 6). **a** Percutaneous transhepatic cholangiography performed following puncture of a left-sided duct with a 22-gauge needle using sonographic guidance. There is dilatation of the left-sided ducts with narrowing of the common hepatic duct by lobular intraductal tumour. The right-sided ducts are not opacified because they are isolated by tumour. **b** Internal-external biliary drainage. An 8-F catheter with numerous side holes has been advanced across the narrowed segment of duct into the duodenum. Following the formation of the distal pigtail, greater catheter stability is achieved compared with external biliary drainage

Results

The clinical characteristics of the patients and the interventional techniques used are summarised in Table 1. Biliary drainage was successful in all six patients.

Biochemical and haematological tests showed a general improvement at 7–9 days (Table 2, Fig. 3). One patient had positive blood culture *(Streptococcus pneumoniae)* and one had positive bile culture *(Acinetobacter* species and *Pseudomonas aeruginosa)* on the day of the EBD. Three patients developed positive bile cultures *(Stenotrophomonas maltophilia* in two and *Staphylococcus aureus* in one) at day 7–9, but there were no positive blood cultures (Table 2). One patient subsequently converted to IEBD developed catheter colonisation with *Candida* species, but did not suffer from systemic fungal infection.

In one patient (case 2), retroperitoneal leakage of contrast medium from the duodenum was detected (Fig. 2 d) after conversion of EBD to bilobar IEBD. There were no other complications of EBD or IEBD.

Discussion

Malignant obstructive jaundice in childhood, although uncommon [2], may be caused by many different types of tumour. The most common is rhabdomyosarcoma of

Table 1. Clinical features and procedures performed in six children and adolescents with malignant obstructive jaundice (*DSCT* desmoplastic small cell tumour, *RMS* rhabdomyosarcoma, *PB* pancreatoblastoma, *HCC* hepatocellular carcinoma, *R* right-sided duct, *L* left-sided duct, *EBD* external biliary drainage, *IEBD* internal-external biliary drainage, *CBD* common bile duct, *TBB* transbiliary forceps biopsy, *RPV* right portal vein, *DOD* dead of disease, *AWD* alive with disease)

Case	Sex/age (years)	Tumor	Type of puncture	R and L ducts isolated	Needle gauge	Catheter type	Further procedure(s)	Duration of EBD or IEBD	Outcome
1	M/17	DSCT	R, dilated	No	21	EBD 8 F	CBD stenting	10 weeks	DOD 18 months
2	M/3	RMS	R, dilated	No	21	EBD 8 F	TBB, bilobar IEBD	3 months	DOD 3 months
3	M/5	PB	L, dilated	No	18	EBD 6 F	IEBD	5 months	DOD 5 months
4	M/14	HCC	L, non-dilated	Yes	18	EBD 6 F	RPV embolization	3 weeks	AWD 16 months
5	M/1	RMS	R, non-dilated	Yes	21	EBD 6 F	IEBD	12 months	DOD 12 months
6	M/16	HCC	L, dilated	Yes	22	IEBD 8 F	None	13 months	AWD 13 months

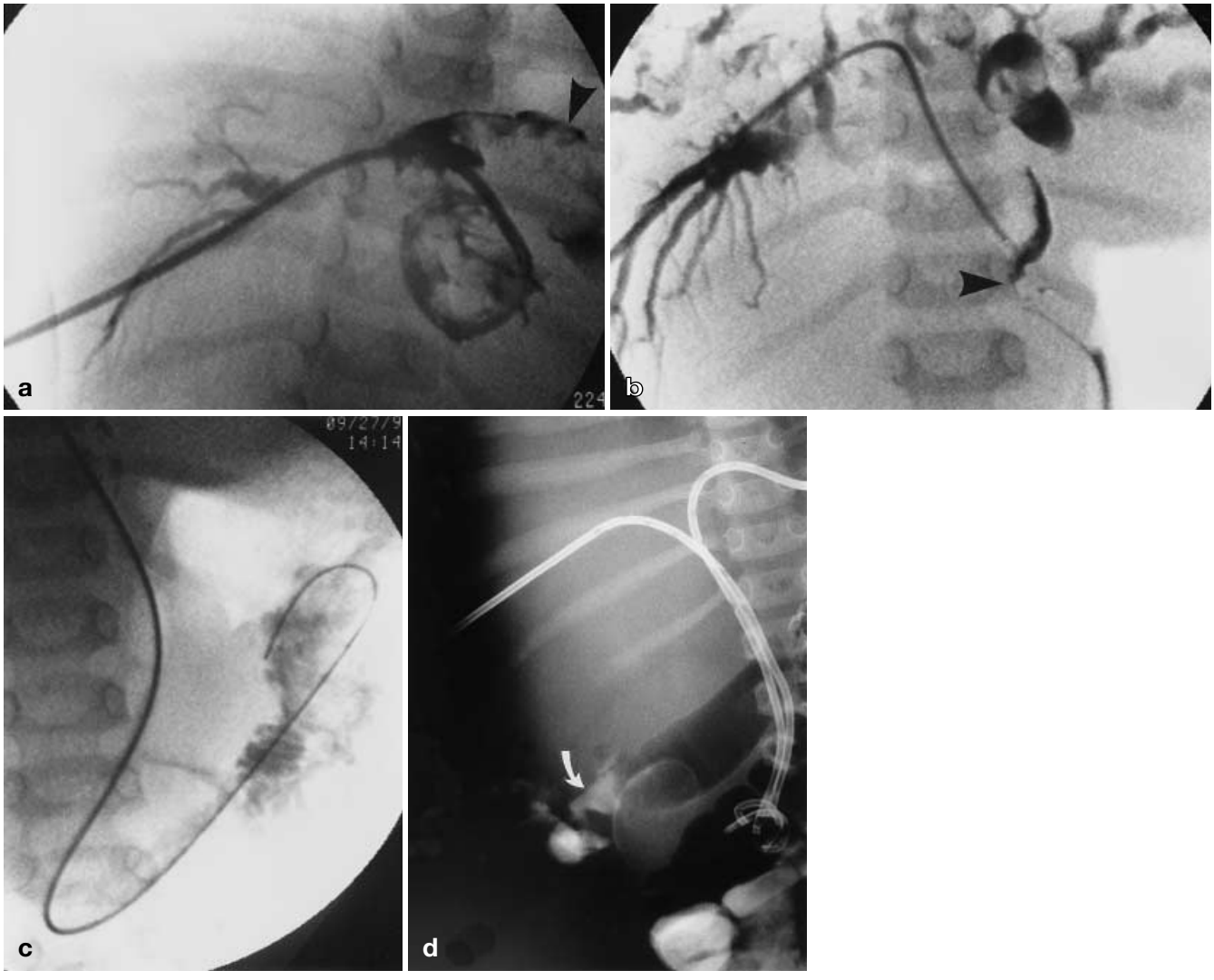

Fig. 2a–d A 3-year-old male with biliary rhabdomyosarcoma (case 2). **a** External biliary drainage. An 8-F pigtail catheter has been positioned above the level of the obstruction. There is extensive lobulated intraductal tumour (*arrowhead*) with no flow of contrast medium into the duodenum. **b** Conversion to IEBD. Following a period of EBD, a small amount of contrast medium drains to the duodenum. The EBD catheter has been exchanged for an angiographic catheter and a hydrophilic guidewire is being used to probe the upper end of the obstruction (*arrowhead*). **c** The catheter has been advanced across the obstruction and an Amplatz super stiff guidewire has been passed into the duodenum prior to insertion of the IEBD catheter. **d** Bilobar internal-external biliary drainage. Additional side-holes have been cut in 8-F nephrostomy catheters to allow internal drainage of bile. There is retroperitoneal contrast medium (*arrow*) resulting from leakage from the duodenum, which was extensively invaded by tumour

the bile ducts or pancreas [3–6]. Neuroblastoma [2, 7–9], Langerhans' cell histiocytosis [2, 10], non-Hodgkin's lymphoma [2, 11], desmoplastic small-cell tumour [1], neuroendocrine tumours [12–14] and granulocytic sarcoma [2] have also been reported to cause obstructive jaundice in the first two decades of life.

Obstructive jaundice must be relieved for several important reasons [15]. Untreated obstruction may cause liver failure, and consequent complications may be aggravated by the adverse effects of chemotherapy, including hepatic toxicity and thrombocytopenia. Certain chemotherapeutic agents commonly used in the treatment of paediatric malignancies, in particular doxorubicin, are excreted in the bile. For these agents, the presence of unrelieved biliary obstruction may cause problems with dose scheduling. Re-establishment of bile flow to the duodenum (by IEBD or stenting) may also improve absorption of dietary fats. This may be important in children with cancer, who often have signifi-

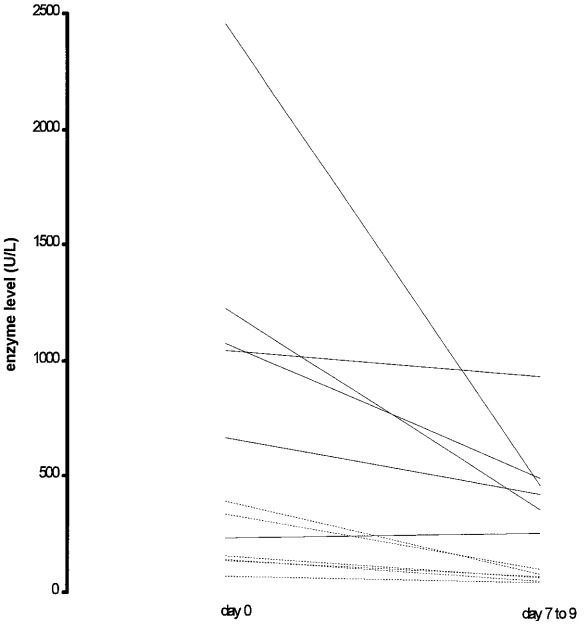

Fig. 3 Change in levels of alkaline phosphatase (*solid lines*) and alanine aminotransferase (*broken lines*) before and 7–9 days after biliary drainage in six children with malignant obstructive jaundice

cant nutritional problems. Patients with malignant tumours that are immediately resectable, or characteristically show rapid shrinkage in response to therapy, for example some forms of non-Hodgkin's lymphoma [16], may not require EBD. In most patients, however, some procedure aimed at relieving biliary obstruction is indicated.

There has been brief mention of the use of EBD in children with malignant obstructive jaundice in the literature [1–3]. IEBD has been performed in a child with benign haemangioendothelioma of the pancreas [17]. There has been no detailed review of the indications for these procedures or their results.

Technical success was achieved in all our patients. Two patients had nondilated ducts. In one, duct dilatation had been observed at diagnostic sonography. In the interval between sonography and EBD, there had been rupture of an intrahepatic duct, with formation of a small biloma. The other patient had previously undergone insertion of a nasobiliary catheter to alleviate the obstruction.

Percutaneous biliary drainage was successful in relieving obstructive jaundice in all our patients, with reductions in bilirubin and liver enzyme levels observed at day 7–9 (Table 2, Fig. 3). In one patient with a previously inserted nasobiliary drain, the alkaline phosphatase level remained within the normal range. In one patient, the prothrombin time was prolonged at day 7–9, and no definite cause for this was identified. White blood cell counts (Table 2) decreased in four patients, probably as a result of chemotherapy.

The organisms cultured from the EBD at day 7–9 were unusual biliary pathogens. *Stenotrophomonas maltophilia* colonisation occurred in two patients, although neither developed clinically significant infection. Cholangitis due to this organism has only been reported following biliary tract instrumentation in patients with malignant obstructive jaundice [18]. In the presence of *Stenotrophomonas maltophilia* colonisation it may be appropriate to use trimethoprim-sulfamethoxazole and ticarcillin/clavulanate as prophylactic antibiotic cover when biliary catheters are exchanged [18]. We suspect that the *Staphylococcus aureus* cultured from bile in one patient was introduced into the biliary tree by his previous nasobiliary drain. Prior to EBD, he had suffered an episode of septic shock following nasobiliary cholangiography. The cholangiogram revealed that segments 5, 6 and 7 were isolated by tumour. The septicaemia may have resulted from infection arising in these undrained segments, or from overdistension of the ducts by contrast. The patient who developed persistent colonisation of his IEBD catheters with *Candida* later in the course of his illness did not develop systemic candidiasis. Despite the immunocompromised status of these patients, catheter-related sepsis does not appear to be a significant problem if adequate biliary drainage can be maintained [18].

One patient developed a duodenal leak after conversion to bilobar IEBD (Fig. 2 d), apparently due to necrosis of his huge duodenal tumour. The perforation may,

Table 2. Hematological and biochemical parameters before (day 0) and after (day 7–9) external biliary drainage in six children and adolescents with malignant obstructive jaundice (*NA* not performed, *WBC* white blood cell count, *ALP* alkaline phosphatase, *ALT* alanine aminotransferase, *AST* aspartate aminotransferase, *PT* prothrombin time)

Case	day 0								day 7–9							
	Bile culture	Blood culture	WBC (nl^{-1})	ALP ($U\ l^{-1}$)	ALT ($U\ l^{-1}$)	AST ($U\ l^{-1}$)	Bilirubin ($\mu mol\ l^{-1}$)	PT (s)	Bile culture	Blood culture	WBC (nl^{-1})	ALP ($U\ l^{-1}$)	ALT ($U\ l^{-1}$)	AST ($U\ l^{-1}$)	Bilirubin ($\mu mol\ l^{-1}$)	PT (s)
1	NA	NA	6.2	1225	160	118	150	NA	NA	NA	3.4	361	67	52	31	12.8
2	NA	–	5.8	1045	339	172	NA	12.8	–	–	3.0	933	105	142	NA	12.8
3	–	+	7.4	2453	137	NA	593	14.4	+	–	1.4	462	72	NA	236	19.4
4	+	–	10.3	234	71	NA	76	11.3	+	–	5.0	254	47	NA	44	11.7
5	–	NA	11.7	667	145	NA	129	11.2	+	–	12.2	425	49	NA	33	12.8
6	–	–	10.7	1076	394	455	147	NA	–	–	13.0	494	80	84	84	NA

however, also have been related to the presence of the two catheters in the duodenum.

A single case of haemorrhage and bile leak following EBD for obstructive jaundice has been reported in a child [2]. It is not clear from this report why this complication occurred and, in particular, whether there was any coagulopathy. Haemorrhage related to EBD was not observed in any of our patients. If significant haemorrhage had occurred, we would have performed hepatic angiography with a view to embolisation [19].

Drainage of both right and left lobes (Fig. 2 d) may be required if the lobar ducts are isolated by tumour, as any inadequately drained part of the biliary tree may become infected. IEBD has certain important advantages over EBD, including catheter stability in small children, and the re-establishment of bile flow to the duodenum. If technically possible, IEBD can be performed as the first drainage procedure. When the obstruction is difficult to cross with a guidewire at the first procedure, it is probably better to insert an EBD catheter, instead of subjecting the child to prolonged manipulation, which may lead to septic complications. Conversion to IEBD is usually easy after a few days of EBD.

Temporary stenting of malignant biliary obstruction presumably reduces the risk of biliary sepsis by removing the potential external portal of infection, but has the disadvantage that further biliary procedures, such as changing the stent or performing cholangiography, will require access to the bile ducts to be re-established. Permanent stenting is not recommended if there is any reasonable prospect of the tumour shrinking significantly in response to therapy [1].

Transbiliary biopsy is sometimes performed in adults [20, 21], but does not appear to have been reported in children. In one of our patients with biliary rhabdomyosarcoma, biopsy was performed at the time of exchange of IEBD catheters. We used flexible and rigid forceps, inserted through a peel-away sheath. Biopsy was performed with cholangiographic guidance and confirmed that viable intraductal tumour was present despite systemic chemotherapy. In combination with PTC and EBD, this would also be a reasonable approach to primary diagnosis in a child with obstructive jaundice due to a tumour with a major intraductal component.

There are several alternatives to EBD in children with malignant obstructive jaundice. Endoscopic retrograde cholangiography (ERC) may be used for diagnosis and intervention. Following ERC, a nasobiliary drain or temporary stent may be inserted [7]. We prefer to perform percutaneous transhepatic cholangiography and EBD for several reasons. Most importantly, diagnostic cholangiography and effective biliary drainage can be achieved with a percutaneous technique whether or not the obstruction can be crossed with a catheter. EBD and IEBD do not require papillotomy and maintain access to the biliary system for future diagnostic and interventional procedures. In particular, it is simple to change the catheter if it becomes infected or occluded [17]. It is unlikely that a randomised trial will be performed to compare techniques based on PTC with ERC, and institutional preference will therefore determine which approach is used.

Surgical cholecystostomy has been advocated for obstructive jaundice due to neuroblastoma [8]. Cholecystostomy is only appropriate when it is known that the tumour does not and will not involve the common hepatic or intrahepatic ducts. EBD is a less invasive technique and is clearly preferable in most patients. Percutaneous cholecystostomy also appears to have few, if any, advantages over EBD. Surgical biliary-enteric bypass is rarely necessary in children with cancer.

In conclusion, EBD and IEBD are effective in the relief of malignant obstructive jaundice in children and offer important advantages over alternative techniques. Conversion to internal stenting is possible, but should rarely be necessary, as long-term drainage is usually not required. Infection does not seem to be a significant clinical problem, providing adequate drainage of all segmental ducts can be maintained. Bilobar drainage may be required when the right and left ductal systems are isolated by tumour.

References

1. Roebuck DJ, Stanley P, Katz MD, et al (1998) Gastrointestinal hemorrhage due to duodenal erosion by a biliary wallstent. Cardiovasc Intervent Radiol 21: 63–65
2. Chen BW, Chang MH, Lin DT, et al (1989) Extrahepatic biliary obstruction caused by cancer of non-liver origin in children: report of 5 cases. J Formosan Med Assoc 88: 819–823
3. Roebuck DJ, Yang WT, Lam WW, et al (1998) Hepatobiliary rhabdomyosarcoma in children: diagnostic radiology. Pediatr Radiol 28: 101–108
4. Geoffray A, Couanet D, Montagne JP, et al (1987) Ultrasonography and computed tomography for diagnosis and follow-up of biliary duct rhabdomyosarcomas in children. Pediatr Radiol 17: 127–131
5. Schweizer P, Schweizer M, Wehrmann M (1994) Major resection for embryonal rhabdomyosarcoma of the biliary tree. Pediatr Surg Int 9: 268–273
6. Ruymann FB, Raney RB Jr, Crist WM, et al (1985) Rhabdomyosarcoma of the biliary tree in childhood. A report from the Intergroup Rhabdomyosarcoma Study. Cancer 56: 575–581
7. Guelrud M, Mendoza S, Zager A, et al (1989) Biliary stenting in an infant with malignant obstructive jaundice. Gastrointest Endosc 35: 259–261

8. Gow KW, Blair GK, Phillips R, et al (1995) Obstructive jaundice caused by neuroblastoma managed with temporary cholecystostomy tube. J Pediatr Surg 30: 878–882
9. Walsh MT, Shah KJ (1989) Neuroblastoma presenting with obstructive jaundice. Br J Radiol 62: 624–626
10. Takayanagi K, Issa M, Cook RC (1980) Obstructive jaundice in infancy due to compression of the bile ducts by malignant lymph nodes. J Pediatr Surg 15: 343–344
11. Lucas GN, Fernando S (1995) Burkitt's lymphoma presenting with obstructive jaundice. Ceylon Med J 40: 170
12. Bembenek A, Lotterer E, Machens A, et al (1998) Neuroendocrine tumor of the common hepatic duct: a rare cause of extrahepatic jaundice in adolescence. Surgery 123: 712–715
13. Newman K, Vates T, Duffy L, et al (1992) Pancreatoduodenectomy with preservation of the stomach and pylorus: a safe and effective alternative in children. J Pediatr Surg 27: 1334–1335
14. Judge DM, Dickman PS, Trapukdi S (1976) Nonfunctioning argyrophilic tumor (APUDoma) of the hepatic duct. Simplified methods of detecting biogenic amines in tissue. Am J Clin Pathol 66: 40–45
15. Roebuck DJ (1998) Interventional radiology in children with hepatobiliary rhabdomyosarcoma. (letter) Med Pediatr Oncol 31: 187–188
16. Dudgeon DJ, Brower M (1993) Primary chemotherapy for obstructive jaundice caused by intermediate-grade non-Hodgkin lymphoma. Cancer 71: 2813–2816
17. Sauer L, Harrison MIR, Bond SJ, et al (1987) Long-term percutaneous biliary drainage in an infant with hemangioendothelioma. J Pediatr Surg 22: 606–608
18. Papadakis KA, Vartivarian SE, Vassilaki ME, et al (1995) Stenotrophomonas maltophilia: an unusual cause of biliary sepsis. Clin Infect Dis 21: 1032–1034
19. Savader SJ, Trerotola SO, Merine DS, et al (1992) Hemobilia after percutaneous transhepatic biliary drainage: treatment with transcatheter embolotherapy. J Vasc Intervent Radiol 3: 345–352
20. Donald JJ, Fache JS, Burhenne HJ (1993) Percutaneous transluminal biopsy of the biliary tract. Can Assoc Radiol J 44: 185–188
21. Elyaderani MK, Gabriele OF (1980) Brush and forceps biopsy of biliary ducts via percutaneous transhepatic catheterization. Radiology 135: 777–778