PST | PEDIATRIC SURGERY IN TROPICS

SOLID PSEUDOPAPILLARY EPITHELIAL NEOPLASM (SPEN) OF THE PANCREAS

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Keywords

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Abstract

Introduction: Solid pseudopapillary epithelial neoplasms (SPENs) of the pancreas are rare neoplasms, particularly uncommon in children and males. They predominantly affect young females in their second and third decades of life. SPENs are typically indolent in nature and often detected incidentally during physical examination or imaging performed for unrelated reasons. According to the World Health Organization (WHO), SPENs are low grade malignant neoplasms with a favourable prognosis.
Case Report: We report a case of an 8-year-old female presenting with clinical features of obstructive jaundice. CECT scan of the abdomen revealed a solid, heterogeneously enhancing mass in the pancreatic head, associated with dilatation of the intrahepatic, extrahepatic, and pancreatic ducts. The patient underwent a Whipple's procedure without intraoperative complications. Postoperative recovery was unremarkable. Histopathological analysis demonstrated a circumscribed, encapsulated neoplasm with mixed solid and pseudopapillary areas; the solid regions contained uniform cells with capillary sized vessels. Immunohistochemistry confirmed SPEN, and an R_o resection was achieved, with no features of malignancy.
Conclusion: This case underscores the importance of considering SPEN in young children presenting with obstructive jaundice. R_o resection provides definitive treatment with excellent prognosis.

Abbreviations	SPENs: Solid Pseudopapillary Epithelial Neoplasms WHO: World Health Organization CECT: Contrast Enhanced Computed Tomography MRI: Magnetic Resonance Imaging

INTRODUCTION

Solid pseudopapillary epithelial neoplasms (SPENs) of the pancreas are rare neoplasms first described by Frantz in 1959.[1] Historically, they were variably termed solid and cystic pancreatic neoplasms, Frantz tumors, solid and papillary epithelial neoplasms, Hamoudi tumors, and papillary cystic neoplasms.[1] In 1996, the World Health Organization (WHO) reclassified them as solid pseudopapillary neoplasms.[2] Malignant SPENs are defined by angioinvasion, perineural invasion, or deep infiltration of pancreatic parenchyma.[2] Despite this potential, SPENs are considered low grade malignancies due to their generally benign morphology and rare metastasis.[3]

SPENs constitute 1-2% of adult exocrine pancreatic neoplasms and roughly 5% of cystic pancreatic lesions. [4,5] In children, they represent 6-17% of pancreatic neoplasms. [4,5] These neoplasms predominantly affect young women but may also occur in males, children, and older adults. Anatomical distribution differs with age: adults typically develop neoplasms in the pancreatic body or tail, while children more frequently have lesions in the pancreatic head.[5] Most pediatric cases present in adolescence (median 11-14 years), though cases in children as young as 7-8 years are documented.[6-8] Patients may present with abdominal pain, biliary obstruction or gastric outlet obstruction.[9,10] The lesion may also be incidentally detected.[9,10] Rarely, SPENs may be discovered after trauma or spontaneous rupture, highlighting their variable clinical spectrum.

Radiologically, SPENs exhibit mixed solid and cystic features on Contrast Enhanced Computed Tomography (CECT) or Magnetic Resonance Imaging (MRI), with larger neoplasms demonstrating cystic degeneration. [11] Peripheral arterial enhancement and central calcifications are typical, and solid components usually enhance similarly to pancreatic parenchyma. [14] Small SPENs may appear entirely solid. Pediatric SPENs are often large at diagnosis (5-10 cm), reflecting delayed detection. The utility of fine needle aspiration remains uncertain. Complete (R_o) surgical resection yields excellent long-term

outcomes with low recurrence. [3,11,13,15] Surgical approach depends on the location of the neoplasm, ranging from enucleation to distal pancreatectomy or pancreaticoduodenectomy. Most SPENs are grossly encapsulated and non-infiltrative, favoring limited resection to reduce morbidity, though microscopic infiltration is common. Chemotherapy or chemoradiotherapy has a limited role, reported mainly for initially unresectable neoplasms. [11]

Overall, SPENs exhibit distinctive clinical, radiological, and pathological features, and despite an occasionally aggressive behavior, surgical excision is curative in most cases. Their favorable prognosis, even in pediatric populations, underscores the importance of early recognition and complete resection. [3,11,13,14,15,18,19]

CASE REPORT

We present the case of an 8-year-old female with a solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas, alongside a current literature review to provide evidence-based guidance for managing this rare pediatric neoplasm.

The patient presented with a 4-month history of progressive jaundice, dark urine, pale stools, abdominal pain, and pruritus. The family had initially sought care from traditional healers and religious practitioners before presenting to our institution, leading to uncertainty regarding the precise duration of symptoms. There were no constitutional symptoms, and her perinatal history was unremarkable. She had no known chronic illnesses, prior hospital admissions, or family history of malignancy.

Clinical Examination and Laboratory Findings

On examination, she was alert, with normal anthropometrics and a Glasgow Coma Scale of 15/15. She was jaundiced with a low-grade temperature of 37.9°C but all other vital signs were within normal limits. Respiratory and cardiovascular examinations were unremarkable. Abdominal examination revealed epigastric fullness with a palpable, non-tender mass. Urinalysis confirmed bilirubinuria. Laboratory investigations were suggestive of obstructive jaundice with total bilirubin 232 µmol/L, conjugated bilirubin 188 µmol/L, γ-glutamyl

transferase 36 → 96 U/L, alkaline phosphatase 362 U/L, aspartate aminotransferase 77 U/L, alanine transaminase 57 U/L, and lactate dehydrogenase 326 U/L. Albumin was 45 g/dL. Mild pancreatitis was present with an amylase 178 U/L and lipase 230 U/L. Coagulation studies showed INR 2.57 and PTT 36.8s. Tumor markers were within normal limits with a CA 19-9 38 U/mL, AFP <1.3 ng/ml, and β-hCG 2 IU/L. Hepatitis studies were negative. Renal function, electrolytes, glucose, hemoglobin, and platelets were all normal.

Imaging

Chest and abdominal plain films were unremarkable. Abdominal ultrasound (Fig 1) demonstrated a 4.54 × 4.6 cm solid mass in the pancreatic head, with dilated pancreatic (0.32 cm in neck; 0.28 cm in body), dilated common bile duct (1.28 cm), distended gallbladder, and normal liver and spleen. CECT of the abdomen demonstrated a 4.7 × 4.3 × 3.7 cm heterogeneously enhancing pancreatic head mass with a hypodense central core and no calcifications.(Fig 2) Mass effects included dilatation of intrahepatic, extrahepatic and main pancreatic ducts (0.36 cm), partial effacement and lateral displacement of the second part of the duodenum, medial displacement of the superior mesenteric vein, and abutment of the inferior vena cava and right renal vessels without thrombosis. The liver, gallbladder, spleen, and bones appeared normal.

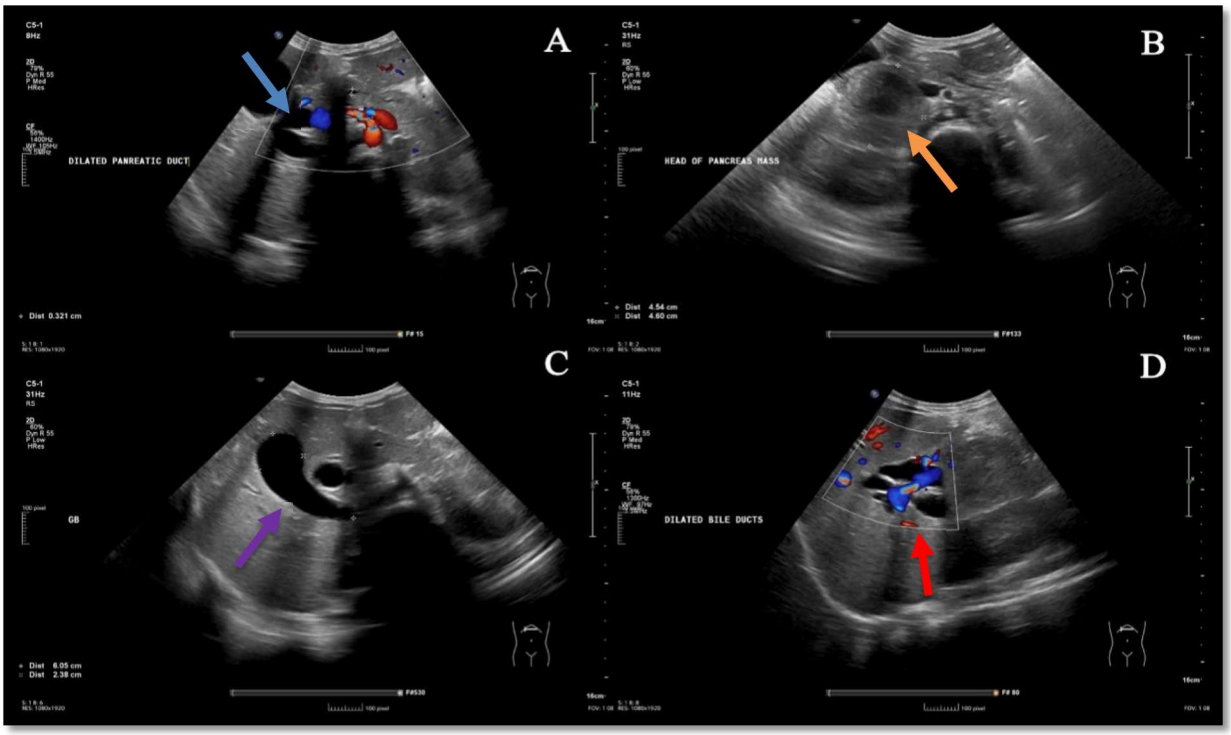

Fig 1. Abdominal ultrasound demonstrating: A) dilated pancreatic duct (blue arrow), B) head of pancreas mass (orange arrow), C) distended gallbladder (purple arrow), D) dilated intrahepatic ducts (red arrow)

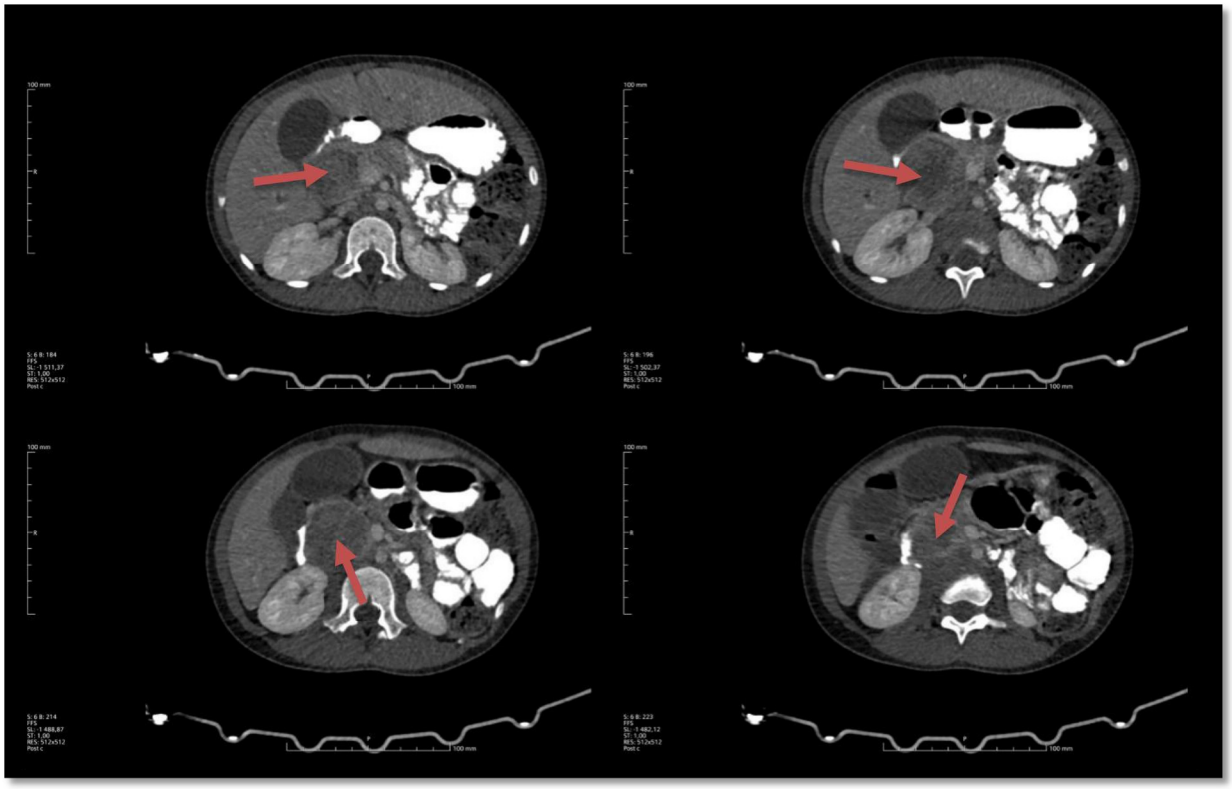

Figure 2. Multiple axial views on the CECT scan of the abdomen; demonstrating the solid head of pancreas mass (red arrows)

Management

The working diagnosis was a pancreatic head mass with secondary mass effect and mild pancreatitis; with differential diagnoses of SPEN and pancreatoblastoma. The patient was started on intravenous fluids and piperacillin/tazobactam. After patient optimization and thorough family counseling about the procedure, an informed consent for surgery was obtained.

Intraoperatively, findings were consistent with preoperative imaging. A Whipple's procedure was performed without complications. (Fig 3 and 4) Postoperatively, the patient was admitted to the pediatric intensive care unit with nasogastric tube, nasojejunal tube and two peritoneal drains in place.

Postoperative Course

The postoperative course was uneventful. Serum amylase and lipase on day 1 were 89 U/L and 49 U/L, respectively, with peritoneal drain amylases showing a downward trend (left drain: 558 → 238 U/L and right drain: 493→ 115 U/L by day 3), thus, not suggestive of a pancreatic fistula. INR (1.28 → 1.16). Liver function tests improved significantly by day 5: total bilirubin 65 → 35 µmol/L, conjugated bilirubin 49 → 29 µmol/L, γ-GT 96 → 31 U/L, ALP 127 → 95 U/L, AST 108 → 57 U/L, ALT 67 → 51 U/L. Hemoglobin remained stable (11.8-12.7 g/dL). She was transferred to the general pediatric surgical ward on day 5, commenced on enteral feeds, and discharged home on day 10. At follow up (1 week, 2 weeks, 1 month, 3 months), she demonstrated complete jaundice resolution and normal liver function.

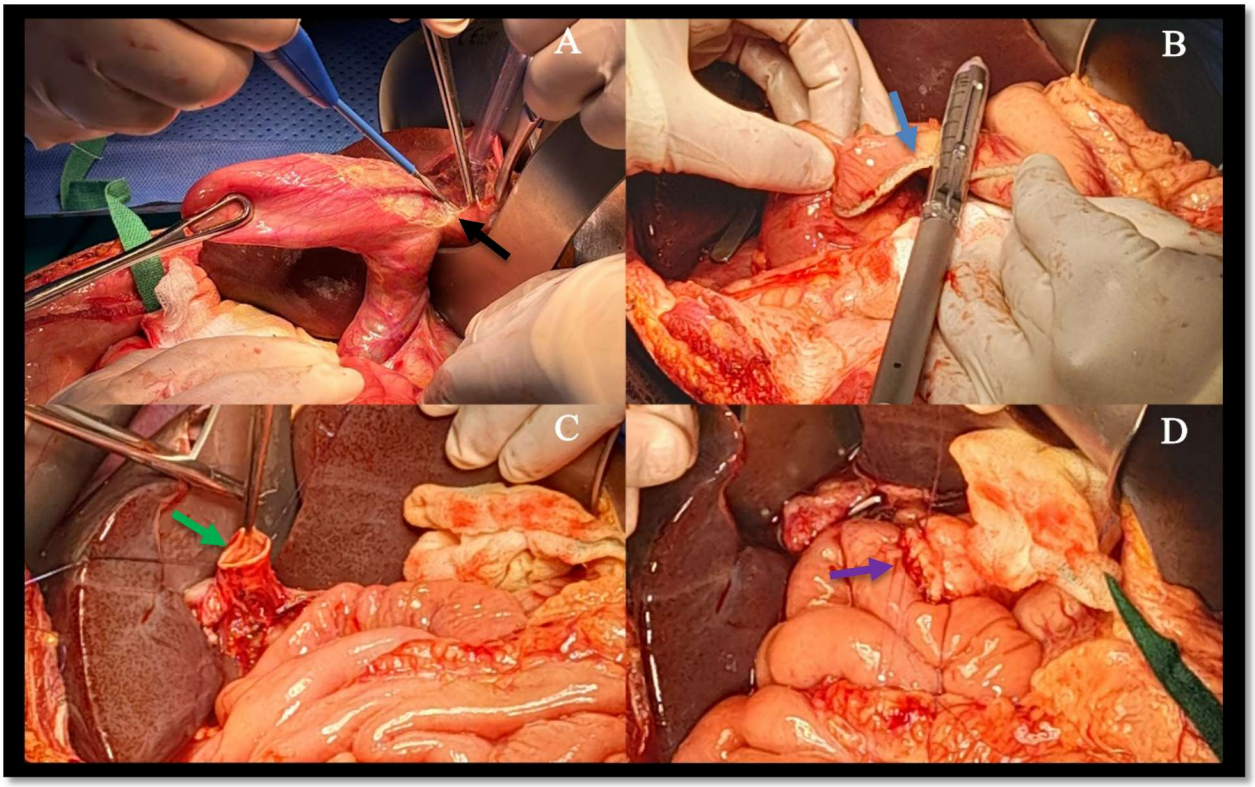

Fig 3 A-D. Demonstrate the Whipple's procedure steps: A) Cholecystectomy (black arrow), B) Duodenectomy, done enbloc with the head of pancreas mass (blue arrow), C) Common hepatic duct stump being prepared for hepaticojejunostomy (green arrow), D) Pancreaticojejunostomy (purple arrow)

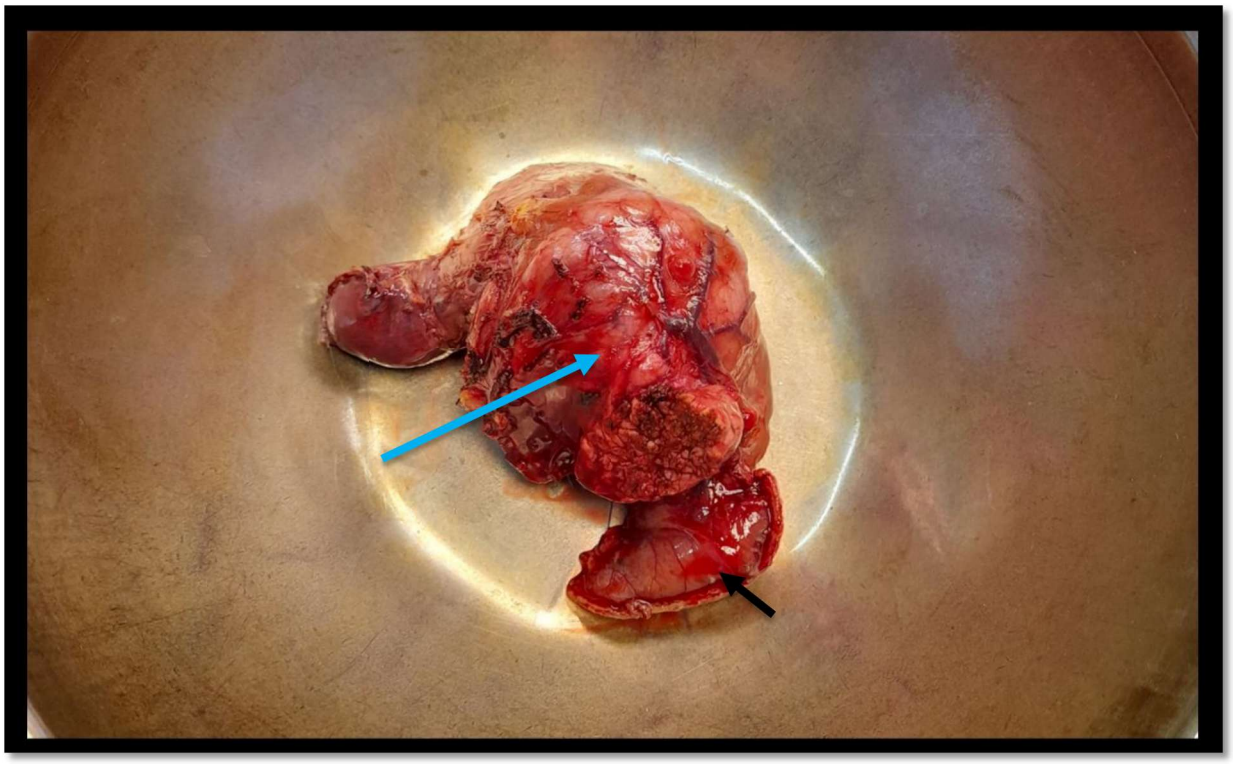

Fig 4. Specimen post resection demonstrating head of pancreas neoplasm (blue arrow) resected enbloc with the duodenum (black arrow)

Histopathology

Microscopy demonstrated a circumscribed, encapsulated neoplasm with mixed solid and pseudopapillary areas. Solid areas contained uniform cells with capillary sized vessels, eosinophilic cytoplasm, fine chromatin, and inconspicuous nucleoli. Pseudopapillae were formed by cells detaching from vessels, creating rosette-like structures. Mitoses were rare, and there was no lymphovascular or perineural invasion. Margins were negative, with the closest serosal margin <1 mm. Immunohistochemistry demonstrated diffuse positivity for CD10 and beta-catenin. These findings confirmed SPEN, and an R_0 resection was achieved.

DISCUSSION

Solid pseudopapillary epithelial neoplasms (SPENs) are rare pancreatic neoplasms. They occur mainly in young women and are very uncommon in children under the age of 10. Our case of an 8-year-old girl adds to this small group and highlights the need to consider SPEN in pediatric pancreatic head masses.

Most pediatric SPENs present in adolescence, with median ages between 11 and 14 years. Younger cases, though rare, have been reported. Symptoms are often vague, including abdominal pain, mass, or nausea. Neoplasms located in the pancreatic head may cause obstructive jaundice or gastric outlet obstruction. Our patient developed progressive jaundice, a less frequent but recognized presentation in this age group.

CECT and MRI scans of the abdomen are the preferred radiological modalities. SPENs typically appear as well-defined masses with solid and cystic components. These features help distinguish them from other pediatric pancreatic neoplasms. In our case, radiological imaging demonstrated a heterogeneously enhancing pancreatic head mass with biliary and pancreatic ductal dilatation, consistent with published reports.

Surgery is the treatment of choice. Surgical options include enucleation, distal pancreatectomy, or pancreaticoduodenectomy, depending on the location of the neoplasm. The key predictor of outcome is a complete (R_o) surgical resection. Our patient underwent a Whipple's procedure, in line with the current recommendations for pancreatic head lesions, and she had an uncomplicated recovery.

Prognosis is excellent after complete (R_o) surgical resection. 5-year survival exceeds 95%, and recurrence or metastasis is rare. However, ongoing surveillance is essential to detect the uncommon late recurrence. Our patients' smooth recovery and expected long-term outcome are consistent with these findings.

This case demonstrates that SPEN, though rare, should be part of the differential diagnosis for pediatric pancreatic masses. It also demonstrates the safety of pancreaticoduodenectomy in children when performed in specialized centers. Reporting such cases helps strengthen the limited data on SPEN in patients younger than 10 years.

CONCLUSION

SPENs are rare pancreatic neoplasms that predominantly affect young females. Clinicians should maintain a high index of suspicion in children presenting with abdominal symptoms and a palpable mass. CECT or MRI scan is recommended for accurate characterization and

surgical planning. Complete (R_o) surgical resection remains the mainstay of treatment. Histopathology and immunohistochemistry are diagnostic, with attention to features of malignancy guiding classification as solid pseudopapillary carcinoma when present. Pediatric SPEN carries a favorable prognosis with low recurrence rates following complete (R_o) surgical excision.

Ethics

Informed consent regarding the personal information about the patient's mother and her child's photographs to be featured in this article. The information will be published without the mother and her child's names attached and every attempt will be made to ensure anonymity. The information may be published in a health journal which is read worldwide.

Conflict of interest

The authors of this article have no financial or personal relationship disclosures that could have any form of influence in this paper.

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