



Case Report

70 Cases of Incomplete Pancreatic Divisum (IPD)-The Usefulness of MRCP Diagnosis and the Safety of Endoscopic Treatments via Duodenal Major and Minor Papilla

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Citation: Tsuji T, Sun G, Shinobi T, Ohishi K, Moriya Y, et al (2024) 70 Cases of Incomplete Pancreatic Divisum (IPD)-The Usefulness of MRCP Diagnosis and the Safety of Endoscopic Treatments via Duodenal Major and Minor Papilla. Ann Case Report. 9: 1967. DOI:10.29011/2574-7754.101967

Received: 02 September 2024, **Accepted:** 05 September 2024, **Published:** 09 September 2024

Abstract

Incomplete Pancreatic Divisum (IPD) is a rare congenital disease caused by maljunction between the ventral duct and the dorsal duct in the 7th week in embryonic stage. In our hospital over the past 10 years, 70 cases of IPD were diagnosed. This is 3.7% (70/1915) of naïve ERP cases during this period (2014.9.~2024.3.). Many classifications were proposed in the literature. We classified them by “modified Hirooka’s classification” into stenotic fusion type 1, 2 (sf 1 7 cases, sf 2 1 case), ansa pancreatica type (0 case) and branch fusion type 1, 2, 3 (bf 1 17 cases, bf 2 0 case, bf 3 44 cases). MRCP was performed in 57 cases, and 20 cases (20/57=35%) were diagnosed by MRCP alone. Finally 49 cases could be correctly diagnosed by MRCP (49/57=86%). Symptomatic cases were treated by endoscopy -13 cases, ESWL -1 case, and ESWL + endoscopy -39 cases. Asymptomatic 14 cases had no therapy.

About the Endoscopic treatment, via major papilla were performed in 19 cases with a technical success rate of 100%, while via minor papilla were performed in 34 cases with a technical success rate of 90% (28/34) without severe complications. In difficult cases, we performed our new endoscopic procedures; rendezvous precut (RP) method and reverse balloon dilation (RBD) method in 8 cases. 81%(43/53) of symptomatic patients had a history of alcohol intake, while 71% (12/17) of asymptomatic case had no alcohol intake. So alcohol intake may make IPD symptomatic.

After endoscopic treatments, the prognosis was good in 48, poor in 4 and 1 had an operation by pain relapse. In 50 calcified cases, ESWL and/or endoscopy were performed more repeatedly than 20 non-calcified cases. EPS is still placed in 39 cases (major 14, minor 25) to maintain the pancreas juice flow and to prevent the papilla occlusion.

Key Words: Incomplete pancreatic divisum, MRCP, Endoscopy

Introduction

In the literature, papers about the diagnosis and the treatment of IPD are few, so we would like to report about MRCP diagnosis and endoscopic treatment in our hospital over the past 10 years. Our aim is to clarify the usefulness of MRCP diagnosis and the safety of endoscopic treatments via the duodenal major and minor papilla.

Methods

70 cases of IPD, experienced over the past 10 years, were enrolled. MRCP diagnoses, endoscopic treatments and their prognoses were evaluated. About the prognosis, good; without irregular pain relapse, controlled with every 2 years EPS replacement, poor; irregular pain relapse uncontrolled with every 2-year EPS replacement

Results

They consisted of 45 males and 25 females, aged 13-90 y/o (mean 63). It was 3.7% (70/1915) of naïve ERP cases in this period. The states of disease were 4 ARP (acute relapsing pancreatitis), 52 CH (chronic pancreatitis), and 14 asymptomatic phase (Table 1).

We classified them by “modified Hirooka’s classification” (Figure 1) into stenotic fusion type1,2, (sf1,sf2), branch fusion type 1,2,3,(bf1,2,3), and ansa pancreatica type. Each number was 7,1,17,0,44 and 0 respectively. One case was unclassified.

53 symptomatic cases consisted of 40 males and 13 females (alcoholic 81%). 6 severe pancreatitis cases with pseudocysts were all calcified alcoholic male cases. While 17 asymptomatic cases consisted of 6 males and 11 females (non-alcoholic 71%) (Table 2). 49 cases were diagnosed by ERP and MRCP, while 20 cases by MRCP alone. Treatment procedures consisted of 39 ESWL+endoscopy (via major papilla 15, via minor papilla 24), 13 endoscopy alone (via major 3, via minor 10), 1 ESWL alone, 1 operation (tail pseudocyst) without medical treatment, and 1 pancreato-duodenectomy after medical treatment and 14 no therapy.

At the first endoscopic treatment, a pancreatic stent (5 or 7Fr. pig tail type) was placed, and 4 months later, it was replaced. Then every 2 years, ERP was performed and if necessary, the stent was replaced. The prognoses were good in 48 cases, however 4 calcified cases required treatments more repeatedly due to stone and pain relapse and a stent is still in place in 39 cases. There was

1 early complication case - duodenal wall perforation and peri-duodenal abscess formation (case E below), while there was no other complication cases such as bleeding, severe pancreatitis. One late complication- stent migration- was experienced. 1 case had an operation (pancreato-duodenectomy) after medical treatment. 3 cases were unsuccessful in endoscopic therapy. There were no cases of cancer occurrence or death after treatments.

Case presentation

A. 79 y/o m bf 3-ipmn(+) and 72 y/o m sf 1 – asymptomatic. diagnosed by MRCP alone (Figure 2).

B. 30 y/o m bf 3: EPST of minor papilla, balloon dilation and EPS (Figure 3).

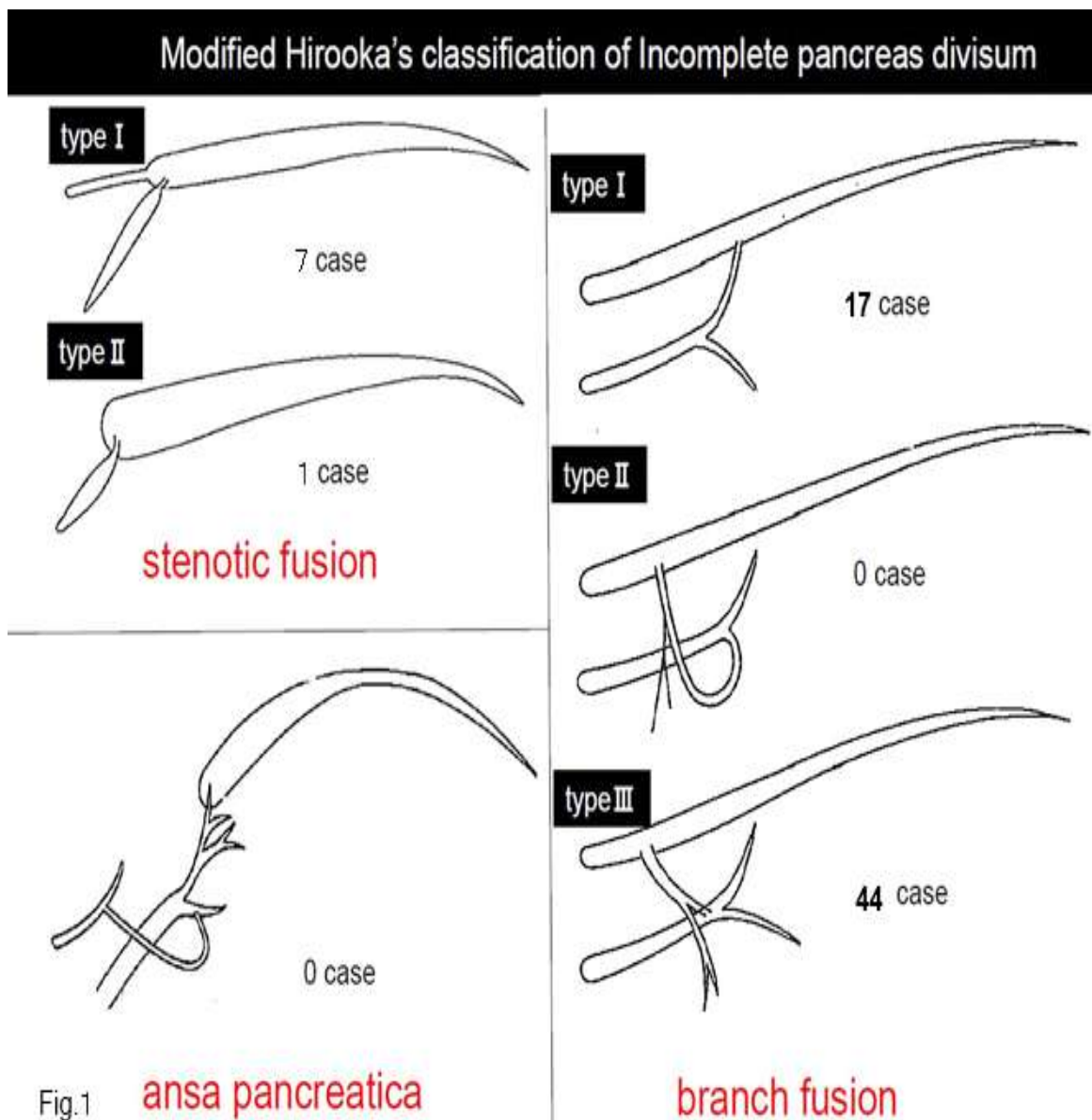
C. Rendezvous pre-cut method: 56 y/o m bf 3: The guidewire, inserted through the major papilla, came out into the duodenum via the minor papilla. Along this guidewire, the minor papilla was cut by the needle type papillotome and the catheter was inserted into the minor papilla, then EPS was placed. This is our original procedure, a variant of the rendezvous method (Figure 4).

D. Reverse Balloon Dilation Method: 13 y/o f bf 3: She entered into our hospital complaining of recurrent epigastralgia. The guidewire, inserted into the major papilla, came out via Wirsung’s duct, connecting branch, Santorini’s duct and minor papilla into the duodenum. The minor papilla was cut by needle type papillotome (rendezvous pre-cut method), then a balloon catheter was inserted along the guidewire and the minor papilla was dilated from the reverse direction by a 4mm dilation balloon, then EPS could be placed into the dorsal duct (Figures 5,6).

E. Reverse Balloon Dilation Method: duodenal wall perforation case 78 y/o f bf 3. When the catheter, proceeded into the duodenum lumen via the minor papilla under the guidewire, injury of the duodenal wall at the opposite side and made peri-duodenal abscess. Percutaneous drainage was performed, then cured. In this method, deep guidewire insertion into the duodenum via the minor papilla is necessary to prevent duodenal wall perforation by catheter (Figure 7).

We had 7 cases combined with IPMN- One case was dorsal duct type IPMN. Minor papilla, located in the duodenal diverticulum, was the drainage route of mucin (Figures 8,9).

Two young females were treated - one was case D above, and the other was a 13 y/o (bf 3) combined with duodenal membranous occlusion.



70 cases of incomplete pancreas divisum (IPD)

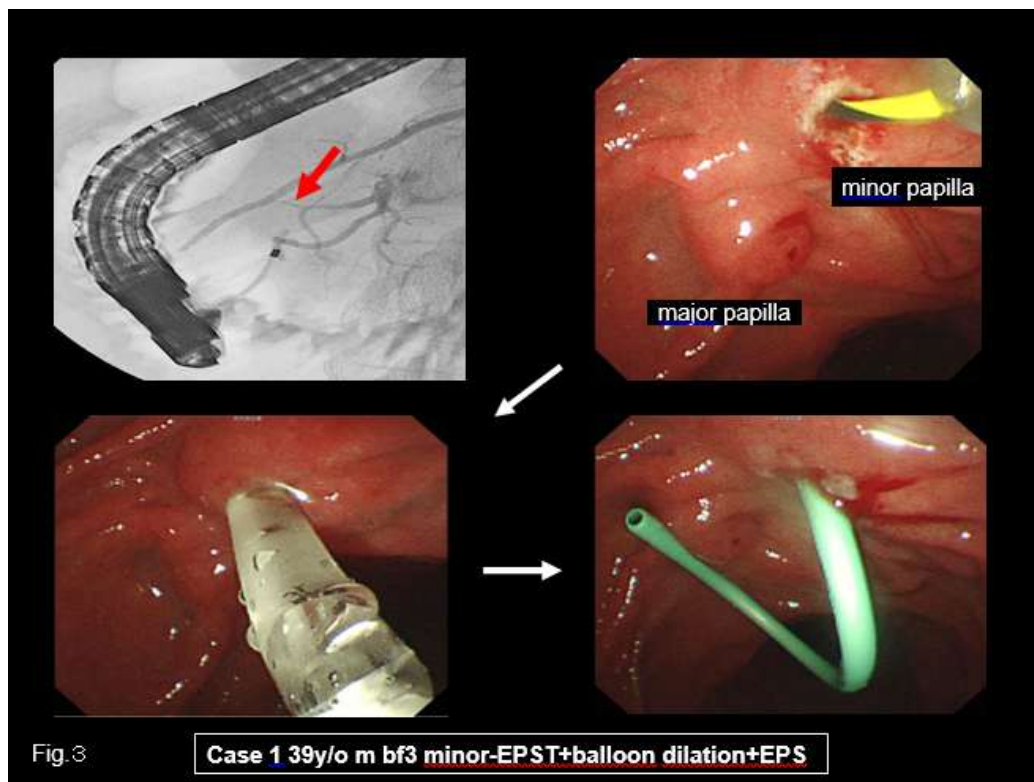
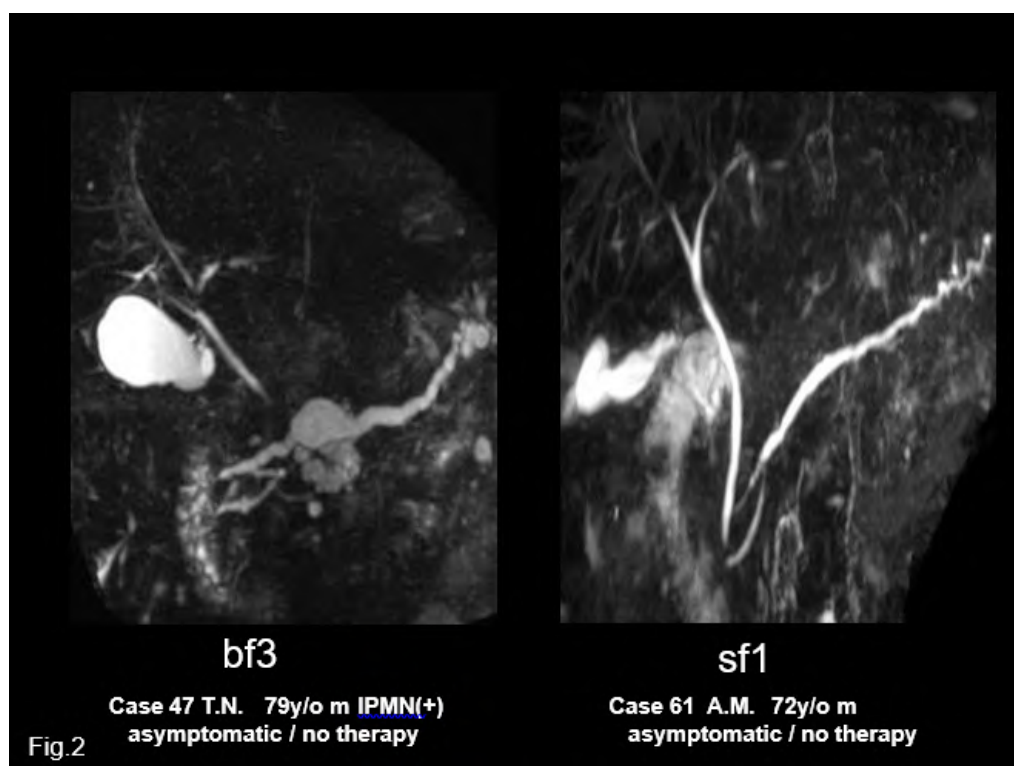
M45 F25 13~90y/o (mean 63) 70/1915 (naive ERCP) = 3.7%
(2014. 4.1, 2023.3.31)

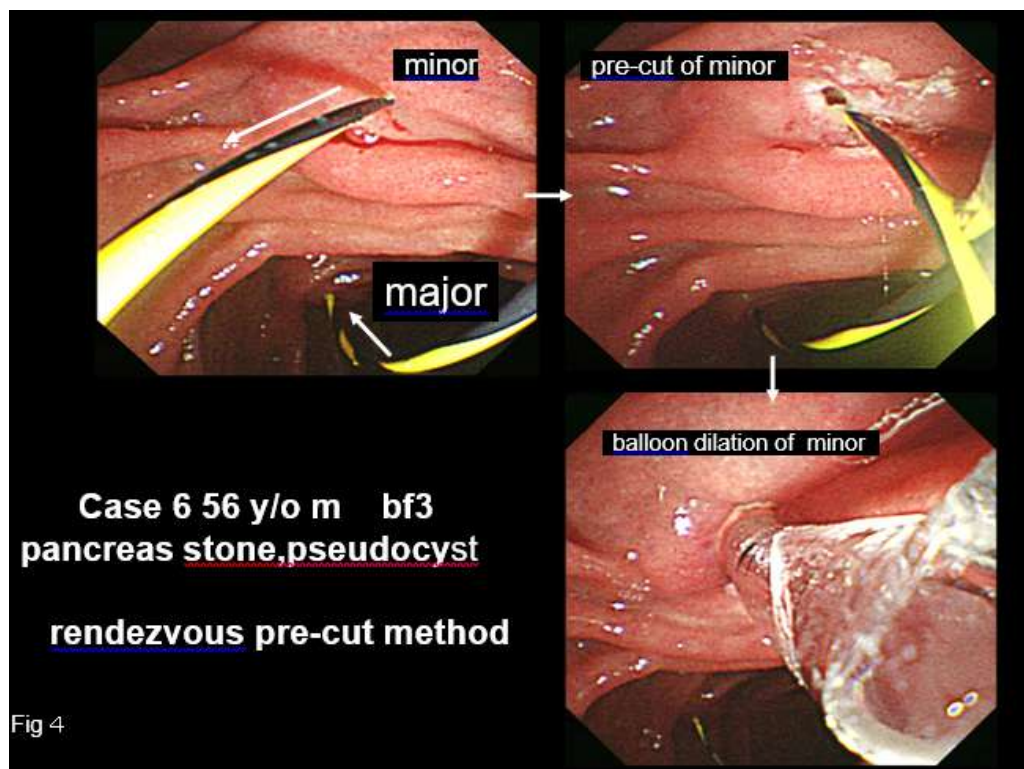
type	stenotic fusion	I 7	II 1	state	ARP	4
	branch fusion	I 17	II 0	III 44	CH	52
	ansa pancreatica		0		asympto.	14
	unclassified		1			
diag.	ERP	50				
	MRCP alone	20				
Treat.	ESWL+ endo (via major 15, via minor 24)	39				
	endo alone (via major 3, via minor 10)	13				
	ESWL alone	1				
	primarily ope	2				
	ESWL+ endo(via major) + ope.	1				
	no therapy	14				
Prognosis after endo.therapy		53				
	Goon course	48				
	Pain relapsing— re-treatment	4				
	Operation	1				
Unsuccessful endo.therapy		3				

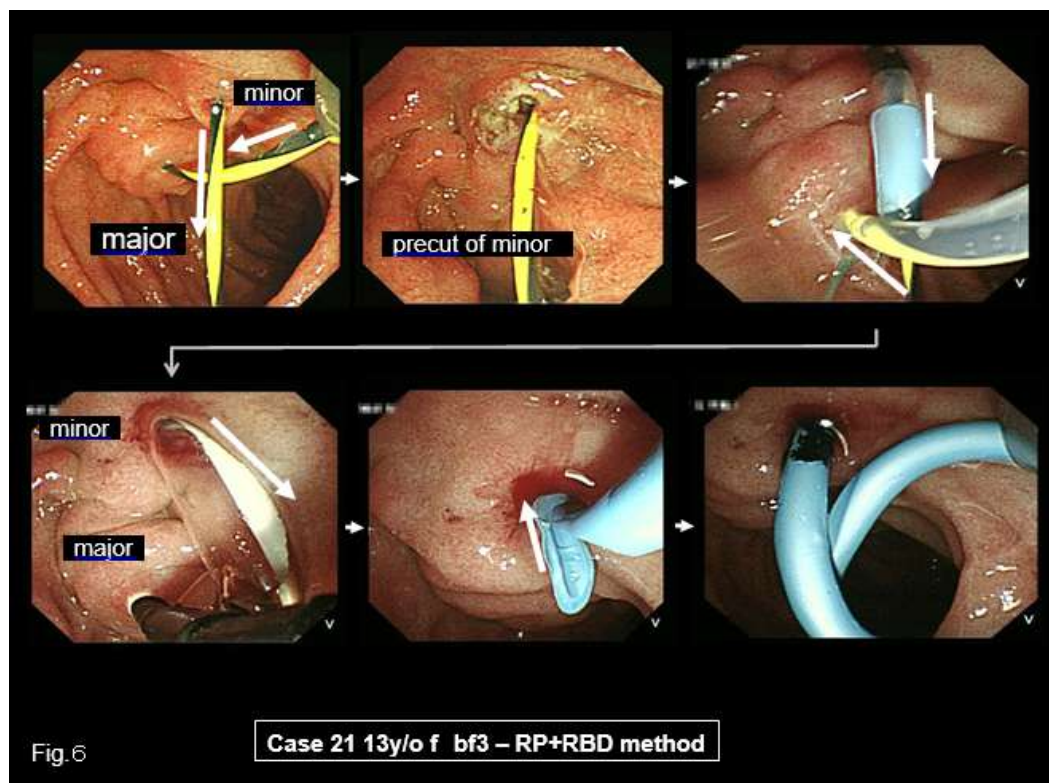
Table 1

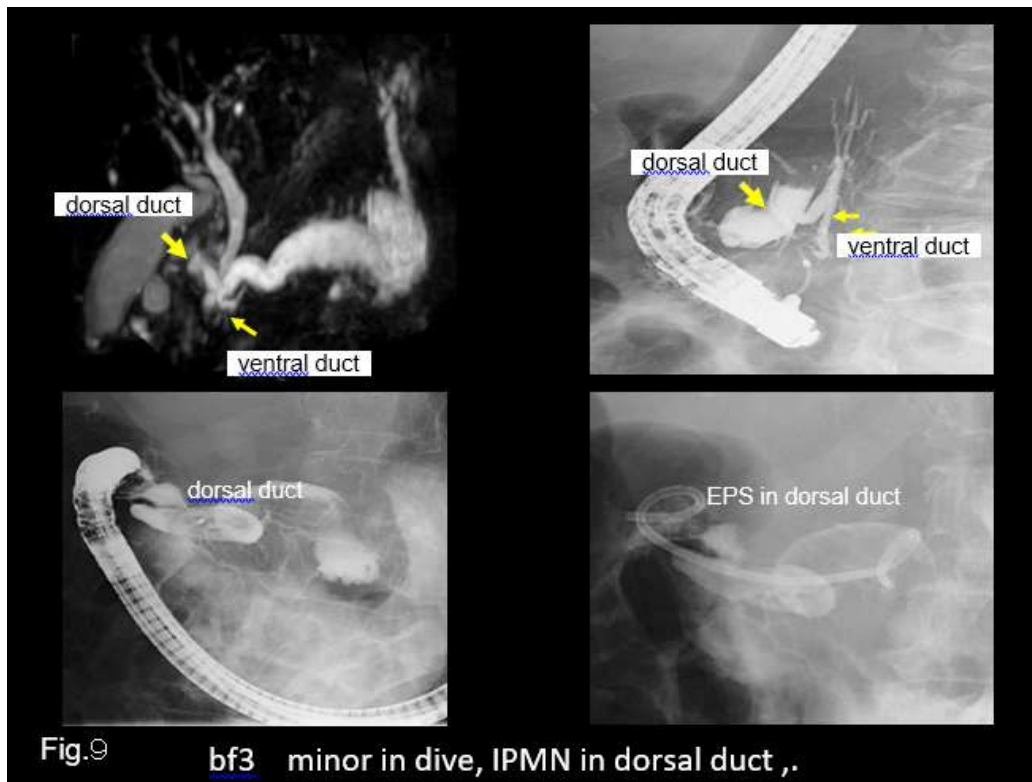
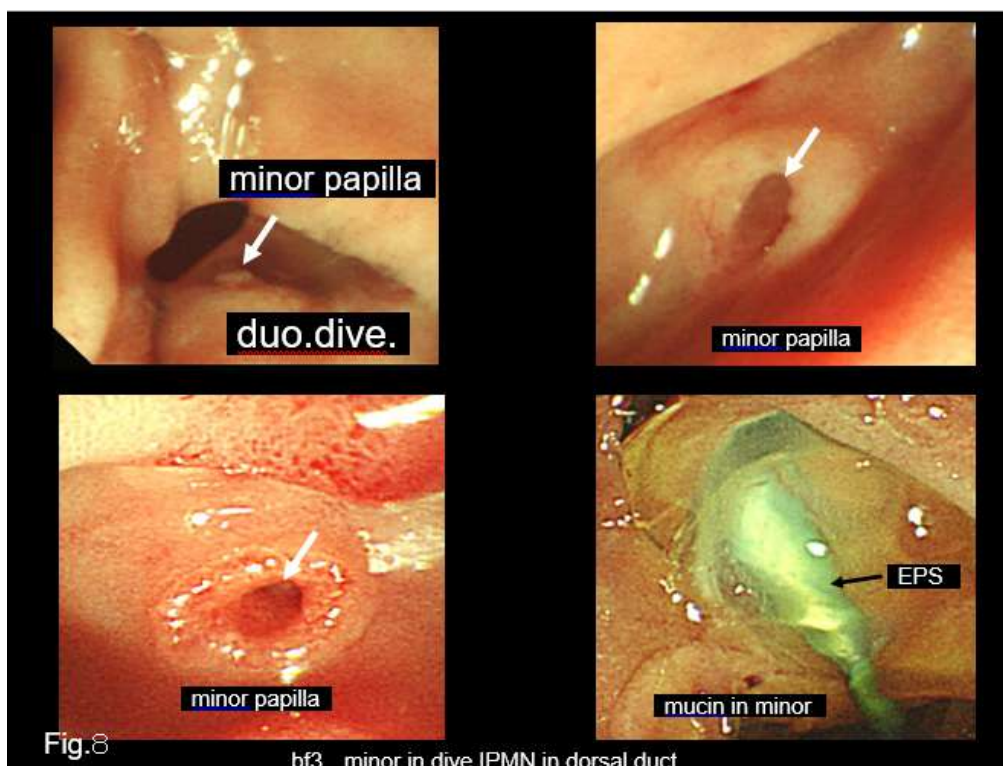
pain (+) 53		pain (−) 17	
M 40 F 13		M 6 F 11	
alcho. (+)			
stone (+)	39		3
stone (−)	4		2
alcho. (−)			
stone (+)	7		1
stone (−)	3		11
43/53 = 81%	alcho. (+)	12/17=71%	alcho. (−)

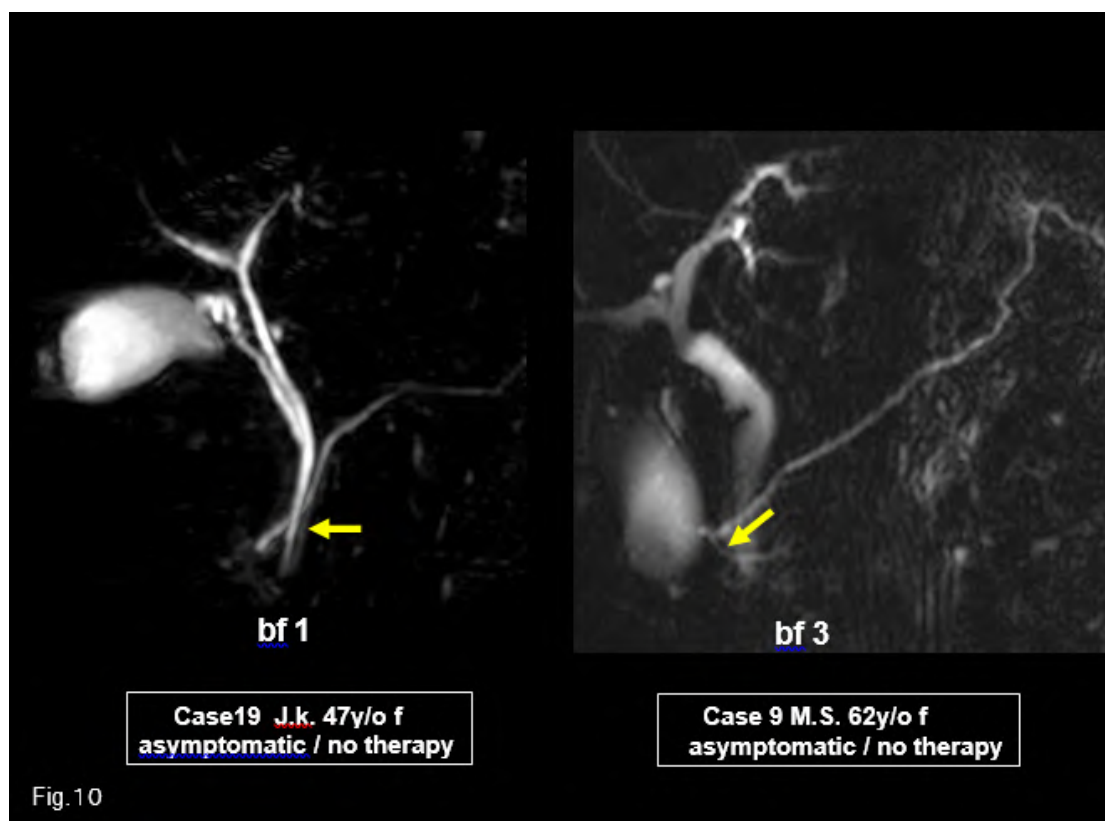
Table 2











Discussion

About the prevalence of IPD in the western countries, it is very low with a reported incidence of 0.13—0.9 % [1-5], on the other hands, it is much high in the recent reports from Japan and Korea [1,6].

We have no data of IPD prevalence in general Japanese population; however, 3.7% (70/1915) of naïve ERP cases were IPD in our hospital over the past 10 years. The mean age in our series was 64.6 y/o except the two young female. Kamisawa reported that the prevalence of IPD was 1.3% (41/3210) in their ERCP series and the mean age was relatively high [1,6].

In the literature, many types of IPD classification were suggested [7,8]. In this paper, we proposed a new type one -“modified Hirooka’s classification”. By this, we classified them into sf1,sf2, bf1,2,3,and ansa pancreatica type. 1 case could not be classified. In the near future, after many reports of IPD, another new useful classification will be proposed. In our reports, there is no bf 2 type, so we suppose some deviations in our diagnosis.

Recently usefulness of MRCP is reported in diagnosis of IPD [9,10]. In our series, MRCP was performed in 57 cases-49 cases (89%) could be diagnosed correctly, and 20 cases were diagnosed by MRCP alone (Figure 10). More IPD cases will be reported

by MRCP diagnosis in the near future. We had no case of cancer occurrence. Kamisawa reported that the cancer occurrence rate was 9% (4/44) in complete and incomplete divisum, mainly in dorsal duct [6,11].

About the treatment, medical conservative therapies were recommended in early stages [12-15] of CPD (complete pancreatic divisum) and IPD. Formally in the symptomatic stage, surgical therapy was performed. In some reports, the therapeutic effects of surgery and endoscopy are almost equal, however, in other reports the effect of the former is superior to that of the latter [16]. In many reports, patients of ARP stage had better results than in CH stage after treatment [17].

Nowadays endoscopic procedures are the preferred choice for CPD and IPD therapy. Cotton reported minor papilla sphincterotomy in CPD [18], and Jacob reported minor papilla sphincterotomy in CPD [19]. Since then, many new techniques (wire-guided minor papilla sphincterotomy, pre-cut method, needle knife cut method, balloon dilation method) were developed and good results were reported by many authors [20-30]. New therapeutic procedures have been reported by Chavan; reverse sphincterotomy of the minor papilla, and by Artifon; reverse minor papilla balloon dilation without EPS [31,32]. From 2017, we have introduced and reported about the new therapeutic procedures for the CPD

and IPD therapy: pre-cut method, balloon dilation method, free-hand method, rendezvous method, rendezvous pre-cut method and reverse balloon dilation method. We reported therapeutic success rates of CPD and IPD via minor papilla was 97% (131/135) and 94% (33/36) respectively without severe complications [33-36]. However, we had to exchange EPS repeatedly and still now EPS is in place in 39 cases (major 14, minor 25). We suppose that the transient metallic stent may be useful in such cases.

Like other author's opinion, we think that the congenital dysfunction of the minor papilla and some acquired factors (ie. alcohol intake, obesity) may make IPD symptomatic [11]. In this report, 81% of symptomatic cases had alcoholic intake histories, while 71% of asymptomatic cases had no such history [37,38].

Conclusions

In this paper, we proposed a new classification – “modified Hirooka's classification” and reported the usefulness of MRCP diagnosis and the safety of endoscopic treatments. Our new endoscopic procedures-rendezvous pre-cut method and reverse balloon dilation method are safe and useful for IPD treatment. After ESWL and/or endoscopic treatment of IPD, the prognoses were good. Acquired factors (alcohol intake, obesity) may make IPD symptomatic.

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