



# Intraductal Papillary Mucinous Neoplasm of the Pancreas: Need for a Tailored Approach to a Rare Entity

Marina Konaktchieva<sup>1</sup>, Dimitar Penchev<sup>2</sup>, Georgi Popivanov<sup>2</sup>, Lilyana Vladova<sup>3</sup>, Roberto Cirocchi<sup>4</sup>, Marin Penkov<sup>5</sup>, Petko Karagyzov<sup>6</sup>, Ventsislav Mutafchiyski<sup>2</sup>

<sup>1</sup> Department of Gastroenterology, Hepatology, and Transplantology, Military Medical Academy, Sofia, Bulgaria

<sup>2</sup> Department of Surgery, Military Medical Academy, Sofia, Bulgaria

<sup>3</sup> Department of Tumour Morphology, University Hospital for Active Treatment of Oncologic Diseases, Sofia, Bulgaria

<sup>4</sup> Department of Surgical Science, University of Perugia, Perugia, Italy

<sup>5</sup> Department of Diagnostic Imaging, St Ivan Rilski University Hospital, Sofia, Bulgaria

<sup>6</sup> Department of Interventional Gastroenterology, Acibadem City Clinic Tokuda Hospital, Sofia, Bulgaria

**Corresponding author:** Georgi Popivanov, Department of Surgery, Military Medical Academy, 3 St Georgi Sofiyski St., Sofia 1606, Bulgaria; E-mail: gerasimpopivanov@rocketmail.com; Tel.: +359 885 521 241

**Received:** 13 Jan 2021 ♦ **Accepted:** 15 Apr 2021 ♦ **Published:** 31 Dec 2021

**Citation:** Konaktchieva M, Penchev D, Popivanov G, Vladova L, Cirocchi R, Penkov M, Karagyzov P, Mutafchiyski V. Intraductal papillary mucinous neoplasm of the pancreas: need for a tailored approach to a rare entity. Folia Med (Plovdiv) 2021;63(6):970-6. doi: 10.3897/folmed.63.e63071.

## Abstract

Intraductal papillary mucinous neoplasm (IPMN) of the pancreas is a relatively new entity that has gained increased attention because of its unique features – presence of different subtypes with different malignant potential, biological behavior, and prognosis, higher rates of recurrences and concomitant or metachronous pancreatic duct cancer. It is rare with an incidence of 4 to 5 cases per 100 000. The relative lack of experience significantly hampers decision making for surgery (pancreatic head resection, distal pancreatectomy or enucleation) or follow-up.

Herein we present two cases managed by diametrically different tactic according to the risk stratification – distal pancreatectomy with splenectomy and observation, respectively. An up-to-date literature review on the key points in diagnostics, indications for surgery, the extent of surgery, follow-up, and prognosis is provided.

The tailored approach based on risk stratification is the cornerstone of management. Absolute indications for surgery are the lesions with high-risk stigmata, whereas the worrisome features should be evaluated by endoscopic ultrasound and fine-needle aspiration. Main duct and mixed type are usually referred to surgery, whereas the management of a branch type is more conservative due to the lower rate of invasive cancer. Strict postoperative follow-up is mandatory even in negative resection margins due to a high risk for recurrences and metachronous lesions.

Despite the guidelines, the intraductal papillary mucinous neoplasm remains a major challenge for clinicians and surgeons in the balance the risk/benefit of observation versus resection. Risk stratification plays a key role in decision-making. Future trials need to determine the optimal period of surveillance and the most reliable predictive factors for concomitant pancreatic duct cancer.

## Keywords

follow-up, imaging diagnostic, intra-ductal papillary mucinous neoplasm, pancreas, surgery, tailored approach

## INTRODUCTION

Intraductal papillary mucinous neoplasm (IPMN) is a cystic pancreatic neoplasm characterized by intraductal papillary proliferation of mucin-producing cells. It is a relatively new entity described in 1980, which rapidly focused the scientific attention because of its unique features – the presence of different subtypes with different malignant potential, biological behaviour and prognosis, higher rates of recurrences and synchronous or metachronous pancreatic duct cancer. It is rare with an incidence of 4-5/100 000, accounting for only 1% of all pancreatic tumours and 20%-30% of cystic neoplasm of the pancreas.<sup>1-4</sup> Approximately 5% of the pancreaticoduodenal resections are due to IPMN.<sup>5,6</sup> Over ten years, Lukanova et al. reported 103 operated patients with rare pancreatic neoplasms, 11% of these being IPMN.<sup>7</sup> The delayed or missed diagnosis is associated with malignant transformation and poor prognosis.<sup>6</sup> An important issue is the differential diagnosis with benign pancreatic tumours.<sup>8,9</sup> Another conundrum is the extremely difficult differential diagnosis with synchronous pancreatic ductal adenocarcinoma (PDAC), which occurs in 5.3% of the cases with IPMN.<sup>1,10,11</sup>

The purpose of the study was to illustrate two different approaches and to present the best available evidence thus facilitating the decision making in this rare entity.

## CASE REPORTS

### Case 1

A 67-year-old female was admitted because of moderate abdominal pain located in the upper right quadrant with back pain, nausea, vomiting, and weight loss of 5 kg. Symptoms lasted for several months before the admission. The patient had a medical history of arterial hypertension and a previous appendectomy. Laboratory findings were Hb, 124 g/l; CRP, 6.3 mg/l; alkaline phosphatase, 399 U/l; glucose, 8.8 mmol/l; ALT, 414 U/l; AST, 11 U/l; amylase, 6 U/l; total bilirubin, 11 µmol/l; and direct bilirubin, 2 µmol/l. Computed tomography showed atrophic pancreas and a sharp lesion with calcification in the pancreatic tail measuring 30/32 mm in size. The decision for surgery was taken according to the risk factors: clinical symptoms, main duct tumour with size >3 cm and a presence of a mural nodule.

Intraoperatively, there was a round-shaped soft tumour with a diameter of 4 cm, located in the tail of the pancreas (Fig. 1). The fresh-frozen section of the lymph nodes from the splenic hilum was negative. Laparoscopic distal pancreatectomy with splenectomy was performed with stapler transection of the pancreas at the level of the portal vein (Fig. 2). The pancreatic stump was oversutured with 3/0 prolene. The duration of surgery was 270 minutes.

The patient had an uneventful recovery and was discharged on the 7th postoperative day. The gross pathology

revealed an atrophic pancreas with multiple round-shaped cysts. Histological examination showed atrophy, lipomatosis, and fibrosis of the pancreatic parenchyma, cystic dilatation of the main duct with a dense fibrotic wall. There were papillary projections with gastric, intestinal and pancreaticobiliary morphology with mild dysplasia as in non-invasive IPMN (Figs 3,4).

### Case 2

A 75-year-old female was admitted to hospital complaining of intermittent nausea and slight upper abdominal pain. The physical examination and blood assay were unremarkable except for the amylase level of 180 U/l. The abdominal ultrasound revealed multiple cystic lesions of the pancreas. The CT and MRI showed a finding consistent with branch type-IPMN multiple cystic lesions with non-enhancing wall and without intramural nodules (Figs 5-7). The lesions were assessed as low risk and the patient was scheduled for observation.

## DISCUSSION

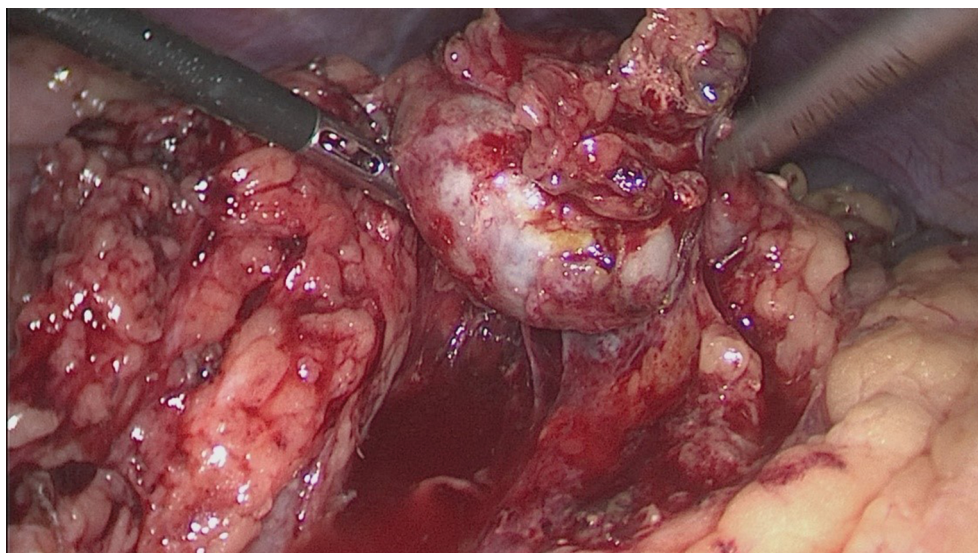
The mastery of IPMN management is the accurate patient selection for surgery, balancing between the unnecessary surgical intervention and overlooking of invasive cancer. Approximately 75% of the patients with IPMN, particularly BD-IPMN, underwent unnecessary surgery.<sup>1</sup> Therefore, the decision to operate or to follow up is pivotal.<sup>12-14</sup>

Based on the morphology, there are three types of IPMN: main pancreatic duct (MD-IPMN, 27%), branch duct (BD-IPMN, 58%) and mixed type (32%).<sup>11</sup> MD-IPMN has significantly higher malignant potential compared to BD-IPMN (43% vs. 18%).<sup>12,13</sup> A more recent survey reported a 23% total rate of invasive cancer in IPMN – 39% in MD, 13% in BD and 32% in mixed type.<sup>11</sup>

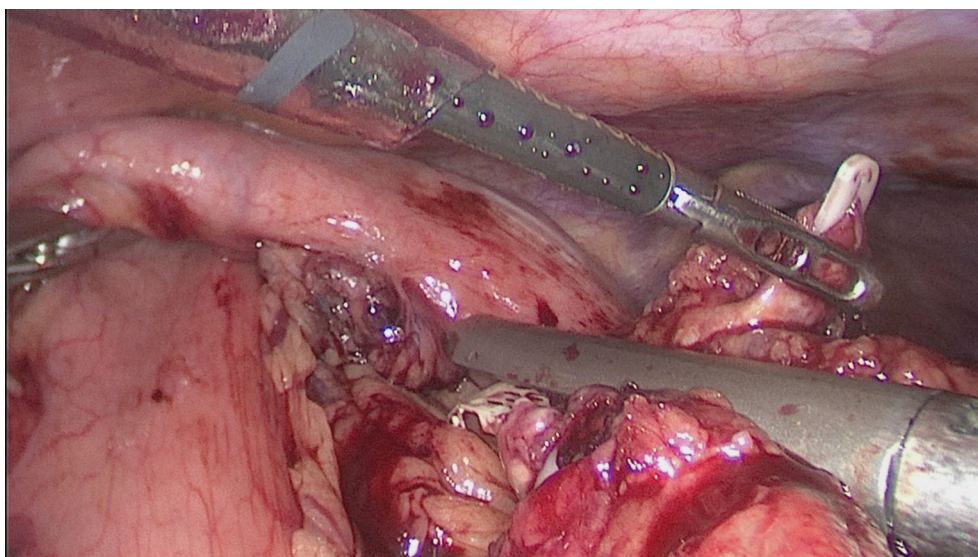
According to the grade of dysplasia, there are four subtypes of IPMN – low, moderate, and high-grade dysplasia, carcinoma in situ and invasive cancer. There are also four histological subtypes with distinct prognosis – gastric, intestinal, hepatobiliary and oncocyte.<sup>12,13</sup>

The revised Fukuoka consensus (2017) divides the patients into two groups.<sup>13</sup> The high-risk group includes obstructive jaundice in a patient with a cyst of the pancreatic head, main pancreatic duct >10 mm and enhancing mural nodule >5 mm. The latter group comprises the so-called “worrisome features” – clinical presentation with pancreatitis, cyst >3 cm, enhancing mural nodule <5 mm, thickened/enhancing cystic wall, the main pancreatic duct 5-9 mm, disconnection of the main pancreatic duct with distal atrophy, lymphadenopathy, increased serum level of CA 19-9, and cyst growth >5 mm/2 years. The mural nodule is the most important predictive factor for cancer, although several studies demonstrated malignant transformation in 9% of the patients without.<sup>13,18,19</sup> On the other hand, Wong et al. reported a 34% incidence of invasive carcinoma in cysts <3 cm, whereas others demonstrated a

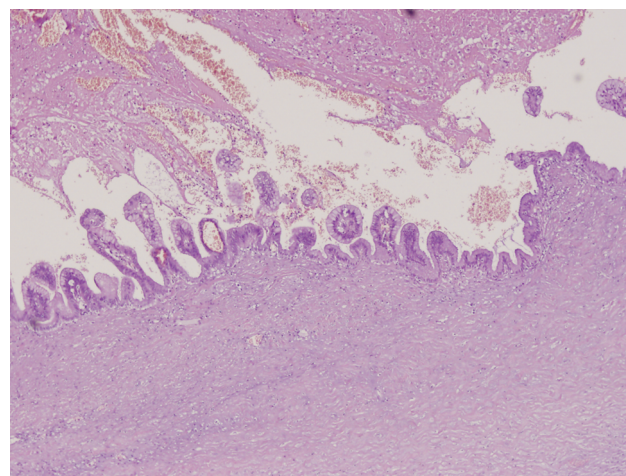
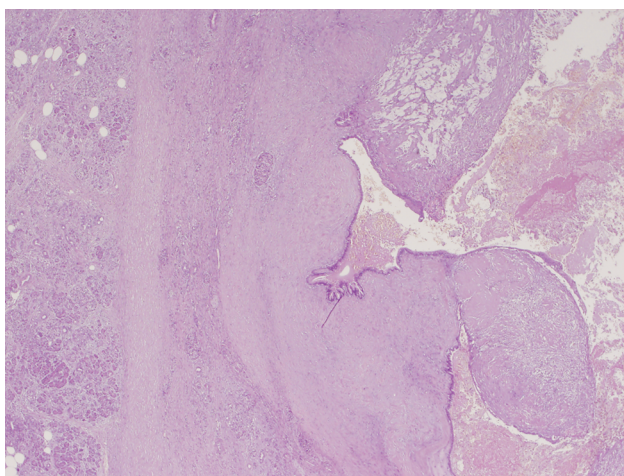




**Figure 1.** Tumour 4 cm in diameter located in the body of the pancreas.

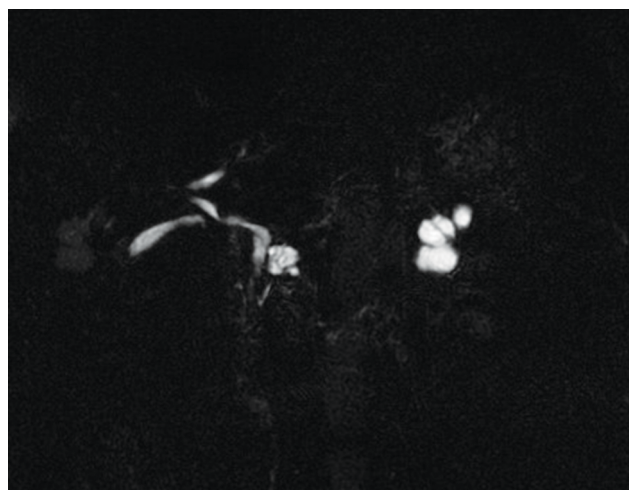
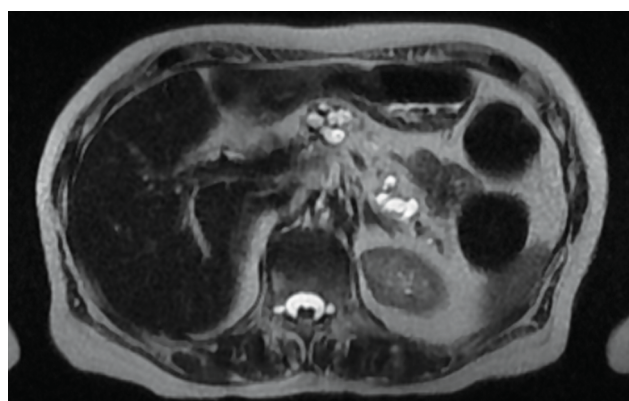
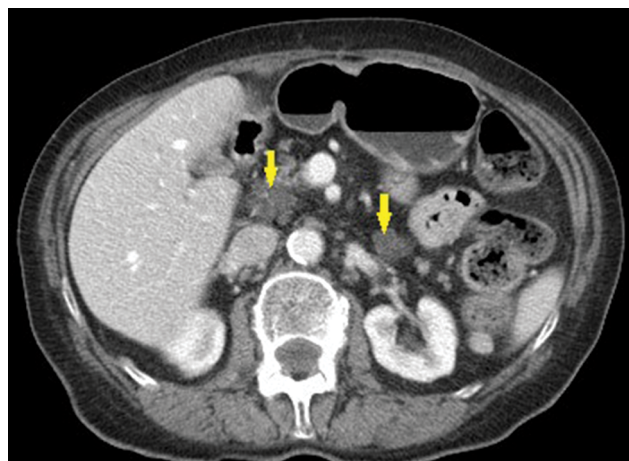


**Figure 2.** Laparoscopic distal pancreatectomy with splenectomy – resection margin at the level of the portal vein.



**Figures 3, 4.** Non-invasive IPMN, mixed type (Hematoxylin-eosin, 2× and 4×).





**Figures 5-7.** CT and MRI and MRCP of branch type IPMN.

59% rate of malignant IPMN in the main duct 5-9 mm.<sup>13,14</sup> These results are an example of lower specificity and sensitivity of all well-known risk factors to predict malignant transformation.<sup>15-17</sup>

The precise preoperative assessment of the malignancy risk is of paramount importance for correct decision-making. MRI/MRCP and CT have a sensitivity of 87% and 83-95%, respectively (Figs 5-7).<sup>20,21</sup> Endoscopic ultrasound (EUS) can precisely evaluate the cystic wall thickening,

communications with the pancreatic ductal system, presence of a mural nodule and allow fine-needle aspiration (FNA) (Fig. 8). EUS can detect mural nodule in 72% of cases without and 98% with contrast enhancement.<sup>20</sup> It is useful for differentiation of IPMN from other cystic lesions through the communication with the pancreatic duct and seems to have higher sensitivity to detect concomitant PDAC than CT and MRI.<sup>21</sup>

The high-risk stigmata are an absolute indication for surgery irrespectively of the type of IPMN. The worrisome characteristic should be further assessed by EUS and FNA – the presence of mural nodule >5 mm, suspicion of main duct involvement or cytology findings suspicious or positive for malignancy are indications for surgery. MD and mixed type are usually referred to surgery, whereas a more conservative approach is used for BD-IPMN because of the lower risk for malignancy (18% vs. 43% in MD). In absence of the worrisome features, CT, MRI or EUS are recommended every 6 to 24 months, although some authors advocate resection in younger patients with cyst >2 cm.<sup>14</sup> Based on the above-mentioned consideration the authors of the Fukuoka consensus stated that “the decision should always be individualized and depends not only



**Figure 8.** US-guided FNA biopsy of high-risk MD-IPMN with the intramural nodule.

on the risk of invasive carcinoma or HGD but also on the patient's life expectancy, comorbidities and cyst location".<sup>14</sup>

On the other hand, some authors suggest that even a 6-month followup is not sufficient for early diagnosis of invasive cancer, so the American Association of Gastroenterology does not recommend an intensive follow-up.<sup>23</sup> However, we and others disagree because strict surveillance is the only chance to diagnose both malignant IPMN and synchronous PDC.<sup>11</sup>

A retrospective study reported no difference in the 5-year survival between resected and non-resected lesions.<sup>24</sup> A recent meta-analysis demonstrated that *"in patients unfit for surgery, IPMN-related mortality among patients with worrisome features and high-risk stigmata is low, and the risk of death from other causes much higher"*.<sup>25</sup> Therefore, we should keep in mind that *"the guidelines are not a religion and surgeons should be thoughtful"* (M. Walsh, World Congress of Surgery, Basel, 2017).

The appropriate surgical procedure is another key step. The standard approach is resection with lymph node dissection because of the high rate of lymph node metastases (54% in invasive IPMN).<sup>26</sup> Pancreaticoduodenal resection is the most common procedure (71%), followed by distal pancreatectomy (12%).<sup>26</sup> Parenchyma-sparing resections such as enucleation, segmental resections (2%) are indicated for single BD-IPMN without malignant transformation or high-risk lesions in multifocal BD-IPMN. The surgeons should keep in mind, however, that they are associated with a higher rate of complications. A total pancreatectomy is indicated in diffuse involvement, particularly in patients with a family history of pancreatic cancer (15%).<sup>18,26,27</sup> The frozen section (FS) has 95% accuracy and is indicated in the case of unclear margins.<sup>28</sup> A large prospective series reported a change in the extent of resection in 30% of the cases leading to an adequate resection in 97%.<sup>29</sup> The main limitation of FS is the so-called "skip" lesions (approximately 10% of IPMN).<sup>28</sup>

The strict follow-up of the operated IPMN patients is mandatory due to the risk for metachronous lesions and a high recurrence rate even in negative resection margins (10-28%).<sup>28,30</sup> Tanaka reported five- and ten-year cumulative incidence of 8% and 12% for all high-risk lesions – 3% and 6% for high-grade dysplasia/invasive IPMN and 4.5% and 6% for PDAC, respectively.<sup>31</sup> A recent large study reported a 12.5% recurrence rate with approximately three-fold increased risk in high-risk BD lesions.<sup>30</sup>

The ten-year survival in non-invasive IPMN is 90% versus 25% in invasive IPMN, the five-year survival in the case of positive nodes is 30% versus 75% without nodal involvement.<sup>32</sup> John Hopkins' experience demonstrated 77% five-year survival for non-invasive IPMN in contrast to 43% for the patients with invasive IPMN.<sup>26</sup> In certain cases, however, the differentiation of IPMN and PDAC can be difficult, because of the similar microscopic appearance and immunohistochemistry.<sup>10,32,33</sup> Invasive cancer of the intestinal type has a better prognosis with a five-year survival of 90% versus 53% for the gastric type.<sup>10</sup> The patients with invasive

IPMN and metastatic lymph nodes have 24% two-year and 0% five-year survival.<sup>26</sup>

There are no specific laboratory markers for IPMN, but several studies showed significantly higher levels of CEA in pancreatic juice in high-risk lesions.<sup>13</sup> The immunohistochemistry for mucin production such as MUC and KL-6 are usually positive in tumours suspected for malignancy with 97% sensitivity and specificity.<sup>3</sup> MUC5AC alone is expressed in gastric type, MUC-1 and MUC5AC are typical for the pancreaticobiliary and oncocytic type, whereas MUC-2 and MUC5AC are specific for the intestinal type.<sup>22,34</sup> A large number of genetic mutations have been studied in IPMN such as BRAF, KRAS, p53, p16, SMAD 4, DPC 4, S100, miR-21, but only GNAS mutation is specific for IPMN. Nevertheless, GNAS mutation has low specificity (60% in high-grade dysplasia).<sup>22</sup>

The exact time interval of surveillance for both resected and non-resected cases remains the most controversial matter.<sup>35</sup> Even after a strict follow-up, some patients develop metastatic PDAC of the pancreatic remnant.<sup>23</sup> We agree with Nakamura et al. that *"further investigation using a prospective protocol with a large number of patients is needed to establish the optimal interval and period of surveillance, and to determine the most reliable risk factors for concomitant PDAC"*.<sup>35,36</sup>

## CONCLUSIONS

The intraductal papillary mucinous neoplasm remains a major challenge for clinicians and surgeons in the balance of the risk/benefit of observation versus resection. Accurate risk stratification plays a key role in the decision-making.

## Acknowledgements

The authors thank Cvetko Petrov for his valuable technical support to the study.

## REFERENCES

1. Baiocchi G, Molino S, Frittoli B, et al. Increased risk of second malignancy in pancreatic intraductal papillary mucinous tumors: Review of the literature. *World J Gastroenterol* 2015; 21(23):7313–9.
2. Olah A, Kollar D. Surgical aspects of intraductal papillary mucinous neoplasms of the pancreas. *Chirurgia* 2015; 5(110):413–7.
3. David A, Kliansky M, Reid-Lombardo K, et al. The clinical relevance of the increasing incidence of intraductal papillary mucinous neoplasm. *Clin Gastroenterol Hepatol* 2012; 10:555–8.
4. Reid-Lombardo K, Sauver J, Li Z, et al. Incidence, prevalence, and management of intraductal papillary mucinous neoplasm in Olmsted County, Minnesota, 1984-2005: a population-based study. *Pancreas* 2008; 37(2):139–44.
5. Cameron J, Riall T, Coleman J, et al. One thousand consecutive pancreaticoduodenectomies. *Ann Surg* 2006; 244:10–5.

6. Shmidt C, White P, Waters J, et al. Intraductal papillary mucinous neoplasms: predictors of malignant and invasive pathology. *Ann Surg* 2007; 246:644–51.
7. Lukanova C, Trichkov C, Mihailov V, et al. Surgical treatment of mucinous cystic neoplasm and intraductal papillary neoplasm of the pancreas. *Khirurgia (Sofia)* 2018; 84(4):148–56.
8. Matsumoto K, Takeda Y, Harada K, et al. Clinical impact of the KL-6 concentration of the pancreatic juice for diagnosing pancreatic masses. *Biomed Res Int* 2015; 2015:528304.
9. Itai Y, Ohashi K, Nagai H, et al. “Ductectatic” mucinous cystadenoma and cystadenocarcinoma of the pancreas. *Radiology* 1986; 161:697–700.
10. Fong Z, Castillo C. Intraductal papillary mucinous adenocarcinoma of the pancreas: clinical outcomes, prognostic factors and the role of adjuvant therapy. *Viszeralmedizin* 2015; 31:43–6.
11. Tanaka M. Intraductal papillary mucinous neoplasm of the pancreas as the main focus for early detection of pancreatic adenocarcinoma. *Pancreas* 2018; 47:544–50.
12. Tanaka M, Adsay V, Chari S, et al. International consensus guidelines 2012 for the management of IPMN and MCN of the pancreas. *Pancreatol* 2012; 12:183–97.
13. Tanaka M, Fernández-del Castillo C, Kamisawa T, et al. Revisions of international consensus Fukuoka guidelines for the management of IPMN of the pancreas. *Pancreatol* 2017; 17(5):738–53.
14. Goh B. International guidelines for the management of pancreatic intraductal papillary mucinous neoplasms. *World J Gastroenterol* 2015; 21(34):9833–7.
15. Wong J, Wever J, Centeno B, et al. High-grade dysplasia and adenocarcinoma are frequent in side-branch intraductal papillary mucinous neoplasm measuring less than 3 cm on endoscopic ultrasound. *J Gastrointest Surg* 2013; 17:78–85.
16. Tanaka M. International consensus on the management of intraductal papillary mucinous neoplasm of the pancreas. *Ann Transl Med* 2015; 3(19):286–95.
17. Xu B, Ding W, Jin D, et al. Decision making for pancreatic resection in patients with intraductal papillary mucinous neoplasms. *World J Gastroenterol* 2013; 19(9):1451–7.
18. Fritz S, Klaus M, Bergmann F, et al. Small (Sendai negative) branch-duct IPMN's: not harmless. *Ann Surg* 2012; 256:313–20.
19. Kim Y, Shin S, Song K, et al. Branch duct intraductal papillary mucinous neoplasm of the pancreas: a single-centre experience with 324 patients who underwent surgical resection. *Korean J Hepatobiliary Pancreat Surg* 2015; 19:113–20.
20. Harima H, Kaino S, Shinoda S, et al. Differential diagnosis of benign and malignant branch duct intraductal papillary mucinous neoplasm using contrast-enhanced endoscopic ultrasonography. *World J Gastroenterol* 2015; 21(20):6252–60.
21. Palmucci S, Trombatore C, Foti P, et al. The utilization of imaging features in the management of intraductal papillary mucinous neoplasms. *Gastroenterol Res Practice* 2014; 2014:765451.
22. Tanaka M. Thirty years of experience with intraductal papillary mucinous neoplasm of the pancreas from discovery to international consensus. *Digestion* 2015; 90:265–72.
23. Tamura K, Ohtsuka T, Ideno N, et al. Unresectable pancreatic ductal adenocarcinoma in the remnant pancreas diagnosed during every-6-month surveillance after resection of branch duct intraductal papillary mucinous neoplasm: a case report. *JOP* 2013; 14:450–3.
24. Wang S, Shyr Y, Chen T, et al. Comparison of resected and non-resected intraductal papillary mucinous neoplasm of the pancreas. *World J Surg* 2005; 29:1650–7.
25. Vanella G, Crippa S, Archibugi L, et al. Meta-analysis of mortality in patients with high-risk intraductal papillary mucinous neoplasms under observation: mortality in patients with high-risk intraductal papillary mucinous neoplasms under observation. *British J Surg* 2018; 105(4):328–38.
26. Sohn T, Yeo C, Cameron J, et al. Intraductal papillary mucinous neoplasm of the pancreas. An updated experience. *Ann Surg* 2004; 239(6):788–99.
27. Käppeli R, Müller S, Hummel B, et al. IPMN: surgical treatment. *Langenbecks Arch Surg* 2013; 398:1029–37.
28. Sauvanet A, Couvelard A, Belghiti J. Role of frozen section assessment for intraductal papillary and mucinous tumor of the pancreas. *World J Gastrointest Surg* 2010; 2(10):352–8.
29. Couvelard A, Sauvanet A, Kianmanesh R, et al. Frozen sectioning of the pancreatic cut surface during resection of intraductal papillary mucinous neoplasms of the pancreas is useful and reliable: a prospective evaluation. *Ann Surg* 2005; 242:774–80.
30. Yan L, Siddiqui A, Laique S, et al. A large multicenter study of recurrence after surgical resection of branch-type intraductal papillary mucinous neoplasm of the pancreas. *Minerva Gastroenterol Dietol* 2017; 63(1):50–4.
31. Tanaka M. Reply to Drs Ball and Bressan. *Pancreas* 2019; 48(2):10.
32. Niedergethmann M, Grützmann R, Hildenbrand R, et al. Outcome of invasive and noninvasive intraductal papillary – mucinous neoplasms of the pancreas (IPMN): a 10-year experience. *World J Surg* 2008; (10):2253–60.
33. Ducreux M, Cuhna A, Caramella C, et al. Cancer of the pancreas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2015; 5:56–68.
34. Paini M, Crippa S, Partelli S, et al. Molecular pathology of intraductal papillary mucinous neoplasms of the pancreas. *World J Gastroenterol* 2014; 20(29):10008–23.
35. Nakamura M, Miyasaka Y, Sadakari Y, et al. Comparison of guidelines for intraductal papillary mucinous neoplasm: What is the next step beyond the current guidelines? *Ann Gastroenterol Surg* 2017; 1:90–8.
36. Tanaka M. Clinical management and surgical decision-making of IPMN of the pancreas: methods and protocols. In: Su G. (eds) *Methods in Molecular biology* (Clifton, N.J.) 2019; 1882:9–22.



# Внутрипротоковое папиллярное муцинозное новообразование поджелудочной железы: необходимость индивидуального подхода к редкой сущности

Марина Конакчиева<sup>1</sup>, Димитр Пенчев<sup>2</sup>, Георги Попиванов<sup>2</sup>, Лиляна Владова<sup>3</sup>, Роберто Чироки<sup>4</sup>, Марин Пенков<sup>5</sup>, Петко Карагьозов<sup>6</sup>, Венцислав Мутафчийски<sup>2</sup>

<sup>1</sup> Отделение гастроэнтерологии, гепатологии и трансплантологии, Военно-медицинская академия, София, Болгария

<sup>2</sup> Отделение хирургии, Военно-медицинская академия, София, Болгария

<sup>3</sup> Отделение онкологической морфологии, УСБАЛ онкологии, София, Болгария

<sup>4</sup> Кафедра хирургических наук, Университет Перуджи, Перуджа, Италия

<sup>5</sup> Кафедра визуализирующей диагностики, УМБАЛ „Св. Иван Рилски“, София, Болгария

<sup>6</sup> Кафедра интервенционной гастроэнтерологии, Аджибадем Сити Клиник УМБАЛ Токуда, София, Болгария

**Адрес для корреспонденции:** Георги Попиванов, Отделение хирургии, Военно-медицинская академия, ул. „Георги Софийски“ № 3, София 1606, Болгария; E-mail: gerasimpopivanov@rocketmail.com; Тел.: +359 885 521 241

**Дата получения:** 13 января 2021 ♦ **Дата приемки:** 15 апреля 2021 ♦ **Дата публикации:** 31 декабря 2021

**Образец цитирования:** Konaktchieva M, Penchev D, Popivanov G, Vladova L, Cirocchi R, Penkov M, Karagyzov P, Mutafchyski V. Intraductal papillary mucinous neoplasm of the pancreas: need for a tailored approach to a rare entity. Folia Med (Plovdiv) 2021;63(6):970-6. doi: 10.3897/folmed.63.e63071.

## Резюме

Внутрипротоковое папиллярное муцинозное новообразование (ВПМН) поджелудочной железы – относительно новый объект, который привлекает всё больше внимания благодаря своим уникальным свойствам – наличию различных подтипов со злокачественным потенциалом, биологическим поведением и прогнозом, более высокой частотой рецидивов и сопутствующей метастатической карциномой поджелудочной железы. Это редкое заболевание – от 4 до 5 случаев на 100 000. Относительное отсутствие опыта значительно ограничивает решение о хирургическом вмешательстве (резекция головки поджелудочной железы, дистальная панкреатэктомия или энуклеация) или последующем наблюдении.

Здесь мы представляем два случая, освоенных с диаметрально противоположной тактикой по оценке риска – дистальная панкреатэктомия со спленэктомией и наблюдение соответственно. Представлен актуальный обзор литературы по ключевым этапам диагностики, показаниям к хирургическому вмешательству, степени хирургического вмешательства, контролю и прогнозу.

Индивидуальный подход, основанный на оценке риска, является краеугольным камнем в управлении этим заболеванием. Абсолютным показанием к операции являются поражения с рубцами высокого риска, а тревожные признаки следует оценивать с помощью эндоскопического ультразвукового исследования и тонкоигольной аспирации. ВПМН основного канала и ВПМН смешанного типа обычно направляются на хирургическое вмешательство, тогда как контроль типа, затрагивающего каналы, является более консервативным из-за более низкой частоты инвазивной карциномы. Строгое послеоперационное наблюдение обязательно даже при отрицательных пределах резекции из-за высокого риска рецидива и метастатических поражений.

Несмотря на предписания, внутрипротоковое папиллярное муцинозное новообразование остаётся серьёзной проблемой для клиницистов и хирургов с точки зрения соотношения риска и пользы наблюдения и резекции. Оценка рисков играет ключевую роль в принятии решений. В будущих экспериментах следует определить оптимальный период наблюдения и наиболее надёжные прогностические факторы для сопутствующего рака протока поджелудочной железы.

## Ключевые слова

наблюдение, визуализирующая диагностика, внутрипротоковое папиллярное муцинозное новообразование, поджелудочная железа, хирургия, индивидуальный подход