# Case 136: Intraductal Papillary Mucinous Tumor (Main Duct Type) of the Pancreas[1]

Ana M. Ferreira, MD
Ana Vaz, MD
Morgado Viana, MD
Paulo Gil-Agostinho, MD
Filipe Caseiro-Alves, PhD

**History**

A 57-year-old man presented with a 3-month history of abdominal pain. Physical examination revealed a large firm palpable mass in the epigastric region. There was no history of alcoholism, trauma, or previous pancreatitis. Laboratory findings were unremarkable. The patient underwent abdominal upright radiography, upper gastrointestinal barium examination, abdominal ultrasonography (US), and abdominal computed tomography (CT).

## Imaging Findings

US (Fig 1) revealed a heterogeneous complex mass that contained cystic and solid areas in the epigastric area. CT revealed an 11.6 × 11.5-cm well-circumscribed mass (Fig 2a) that was localized to the uncinate process of the pancreas and displaced but did not invade the adjacent viscera, namely, the right kidney. After intravenous injection of contrast media (Figs 2b–2d), heterogeneous enhancement of the mass was seen. This enhancement revealed a peripheral enhancing thick rim that surrounded a central nonenhancing low-attenuation area. At a slightly higher level, the main pancreatic duct was markedly dilated distally (Fig 2b) and displayed discrete intraluminal solid enhancing nodules (Fig 2c). The adjacent pancreatic tissue appeared slightly atrophic (Fig 2b). There was no intra- or extrahepatic bile duct dilatation, nor were there any extrapancreatic signs of malignancy. No abnormalities were seen in the rest of the abdomen. Surgery was performed with a Whipple procedure. Pathologic examination of the surgical specimen revealed a yellowish solid tumor (Fig 3). Histologic analysis revealed the papillary component of this huge intraductal papillary mucinous tumor (IPMT) of the pancreas.

## Discussion

IPMT of the pancreas is a pancreatic cystic neoplasm, and it represents about 1%–2% of pancreatic exocrine tu-

**Figure 1**

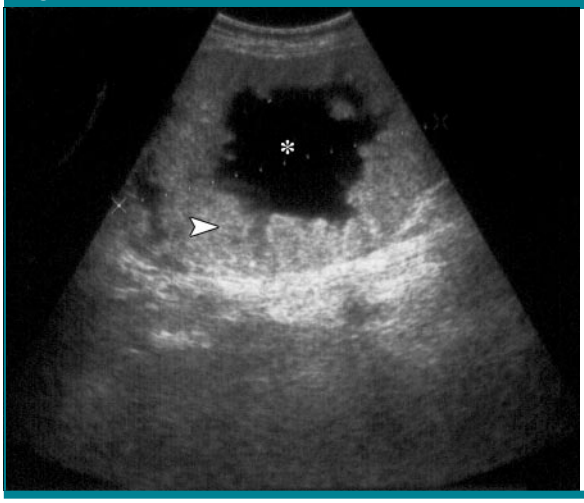

**Figure 1:** Upper abdominal transverse US image shows a heterogeneous complex mass that contains both cystic (*) and solid (arrowhead) areas.

**Published online**
10.1148/radiol.2482031139

**Radiology 2008;** 248:695–698

[1] From the Departments of Radiology (A.M.F., A.V.) and Surgery (M.V.), Hospital Infante D. Pedro-Aveiro, Avenida Artur Ravara, 3814-501 Aveiro, Portugal; and Department of Radiology, Hospital Universidade de Coimbra, Coimbra, Portugal (P.G., F.C.). Received July 31, 2003; revision requested October 21; final revision received June 18, 2005; final version accepted August 5. **Address correspondence to** A.M.F. (e-mail: *aferreira@essua.ua.pt*).

Authors stated no financial relationship to disclose.

mors (1). IPMTs occur with equal frequency in male and female subjects, and the peak time of occurrence is the 6th decade of life, regardless of sex (2). Approximately 75% of IPMTs originate from the main pancreatic duct within the head of the pancreas (3). Imaging of IPMTs is important not only to identify the tumor but also to determine the appropriate treatment strategy in relation to the site and size of the lesion (2). Although the presence of a dominant mass of considerable size (as in the present case) is a rare occurrence, it should not preclude the diagnosis of IPMT.

Imaging findings vary according to the ductal involvement of the disease, which can be divided into three types: *(a)* the main duct type, *(b)* the branch duct type, and *(c)* the combined type (4). In main duct IPMTs, the lesion is usually homogeneous and hypoechoic on US images and hypoattenuating on CT images. Intraductal filling defects can be seen in the main duct. These defects represent mucin globules, papillary projections of the tumor that are hyperechoic on US images, or both (2,5). At CT, unenhanced mucin deposits can be distinguished from contrast-enhanced papillary proliferations (5). Sometimes, it is possible to identify amorphous intraductal calcifications, which are thought to represent calcium deposits within the mucinous collections and which may lead to an incorrect diagnosis of chronic pancreatitis (6). Although Fukushima et al (7) reported 5.0–10.5-cm-diameter IPMTs, large size is an uncommon feature of this tumor.

Branch duct IPMTs, which more frequently are located in the uncinate process, may have a macrocystic or microcystic pattern. The microcystic pattern is characterized by multiple thin septa separating fluid-filled spaces showing US, CT, and magnetic resonance cholangiopancreatography (MRCP) features, such as clustered small cysts with a lobulated contour (2,4). The macrocystic pattern, which is seen more frequently, is characterized by a unilocular or multilocular internal architecture (2). If the tumor is limited to one of the branch ducts, it has a unilocular cystic appearance (4). In later stages, the main duct may also be dilated because of mucin accumulation (2), and severe pancreatic atrophy may follow (4). Demonstration of communication between the lesion and the main duct leads to the correct diagnosis and can occasionally be shown by using thin-section spiral CT (2).

In the combined type of IPMT, both the branch ducts and the main pancreatic duct are involved (4). The duodenal papilla may protrude into the duodenal lumen (4). This finding, which is more frequent in malignant forms (4), is virtually pathognomonic of IPMT (2).

Endoscopically, the ampulla of Vater is often prominent, giving rise to an abundant discharge of mucinous fluid (8). Endoscopic retrograde cholangiopancreatography (ERCP) may be the

**Figure 2**

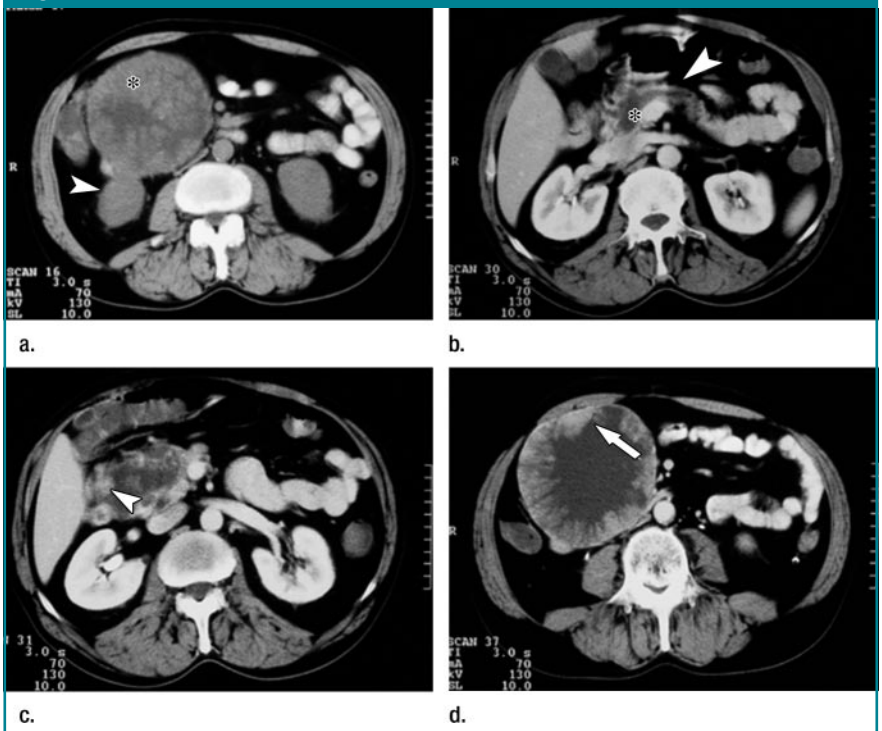


**Figure 2:** Transverse **(a)** unenhanced and **(b–d)** contrast material–enhanced (120 mL, 350 mg of iodine per mL) abdominal CT scans at the level of the palpable mass. In **a**, an 11.6 × 11.5-cm well-circumscribed mass (*) localized at the pancreatic head can be seen. The right kidney (arrowhead) is also visible. In **b**, the main pancreatic duct (*) is slightly dilated, and the adjacent pancreatic tissue (arrowhead) appears slightly atrophic. In **c**, a mass with a peripheral enhancing thick rim surrounding a central low-attenuation area is visible. A nodule (arrowhead) can also be seen. In **d**, there are some discrete intraluminal solid enhancing nodules (arrow).

**Figure 3**

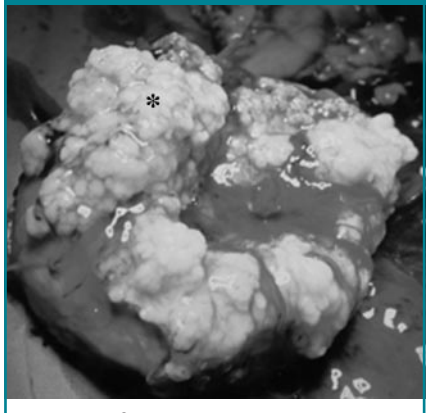

**Figure 3:** Gross solid tumor specimen containing yellowish papillary projections (*) at the periphery.

most direct method with which to diagnose IPMT when it shows the characteristic appearance of mucin discharge from a protruded papilla of Vater (9), the dilated pancreatic duct with intraluminal filling defects due to the presence of mucous plugs (10), and the direct communication of the cystically dilated ductal segment with the main pancreatic duct (2). The dilated duct may lead to a grapelike cluster of cysts with pooling of contrast material, or it may be obstructed by the mucin or the hyperplastic papillary epithelium (10). Sometimes, it demonstrates a fistulous tract leaking mucin between the lesion and the duodenal wall (6). The ERCP data are not constant because mucous plugs may occlude the patency of the ampullary orifice, obviating retrograde opacification by the contrast material (6).

MRCP is more sensitive than ERCP in the detection of cystic dilatation of the side branches (11), and it enables one to avoid the complications associated with ERCP (9). Nevertheless, inspection of a patulous papilla is only possible with use of ERCP (9), as it is superior to MRCP for depicting direct communication of the lesion with the main pancreatic duct (2). Perhaps secretin-enhanced MRCP may have an added value in this regard (2).

Despite its unusually large size, IPMT was a likely diagnosis because it appeared as a cystlike tumor of the pancreatic head in a middle-aged man, with CT features of main duct dilatation containing solid papillary projections. The findings in this patient suggest several diagnoses for a cystic pancreatic mass: *(a)* mucinous cystic neoplasm, *(b)* serous cystadenoma, *(c)* solid and papillary epithelial neoplasm, and *(d)* nonfunctioning islet cell tumor.

Mucinous cystic neoplasms of the pancreas occur in middle-aged women, and the majority of these lesions are located in the pancreatic body or tail (12). They tend to be large; indeed, many are larger than 10 cm (13). They can manifest in the form of abdominal pain, a palpable mass, dyspepsia, anorexia, weight loss, nausea, or vomiting. Symptoms usually result from compression or displacement of neighboring organs and are commonly insidious and of long duration. When symptoms are located in the pancreatic head, they cause early jaundice, which is commonly seen in malignant neoplasms (12). Pathologically, mucinous cystic tumors are encapsulated and multiloculated, with fewer than six cysts being larger than 2 cm in diameter (14). The cystic walls are often thickened, and irregular septa or papillary projections may be seen (14). The amount of stroma varies and, when small, the septa may not be apparent (15). Contrary to IPMTs, mucinous cystic tumors tend to occur in female patients and are generally located in the body or tail of the pancreas, without accompanying ductal dilatation.

In 1978, Compagno and Oertel (16) differentiated serous cystadenoma from mucinous cystic tumor. Serous cystadenomas are almost always benign; malignant forms are exceedingly rare. Benign serous cystadenomas occur predominantly in elderly women without predilection for a particular segment of the pancreatic gland. Benign serous cystadenomas are often an incidental finding or are seen in patients who present with nonspecific clinical features. The median size of these lesions ranges from a few millimeters to many centimeters. The macroscopic structure of these lesions is greatly variable. Although these lesions are characteristically microlacunar, tumors with a mixed structure consisting of a microlacunar core surrounded by cysts larger than 2 cm in diameter, as well as macrolacunar tumors with unilocular cysts usually larger than 2 cm in diameter, have been reported (10). The preponderance of lesions in women, the central calcification, and the microcystic appearance enable one to readily distinguish this entity from IPMT.

A solid and papillary neoplasm is an uncommon low-grade malignancy that occurs predominantly in young women (mean age, 25 years). These tumors are typically large and encapsulated, have a mean size of 9 cm, and are most commonly located in the tail of the pancreas. The internal architecture shows variable proportions of solid and cystic areas, indicating cystic degeneration and necrosis (17,18). Some cases can be exclusively solid, whereas others may be entirely cystic (18). This entity could be ruled out not only because of the tumor location but also because this patient was a 57-year-old man.

Nonfunctioning islet cell tumors should also be included in the differential diagnosis because they are clinically silent until they are large or until they metastasize, and they may grow to be large (3–24 cm in diameter), with cystic change and necrosis (19). However, contrary to IPMTs, nonfunctioning islet cell tumors do not have ductal dilatation and internal enhancing papillary projections as imaging features.

**Acknowledgments:** We thank Abílio Brandão, MD, Department of Pathology, Hospital Infante D. Pedro-Aveiro, and Augusta Cipriano, MD, Department of Pathology, Hospital Universidade Coimbra, for having facilitated and labeled the histologic images.

## References

1. Taouli B, Vilgrain V, Vullierme MP, et al. Intraductal papillary mucinous tumors of the pancreas: helical CT with histopathologic correlation. Radiology 2000;217(3):757–764.

2. Procacci C, Megibow A, Carbognin G, et al. Intraductal papillary mucinous tumor of the pancreas: a pictorial essay. RadioGraphics 1999;19(6):1447–1463.

3. Klöppel G. Clinicopathologic view of intraductal papillary-mucinous tumor of the pancreas. Hepatogastroenterology 1998;45(24): 1981–1985.

4. Lim JH, Lee G, Oh YL. Radiologic spectrum of intraductal papillary mucinous tumor of the pancreas. RadioGraphics 2001;21(2):323–337.

5. Procacci C, Schenal G, Chiara ED, Fuini A, Guarise A. Intraductal papillary mucinous tumors: imaging. In: Procacci C, Megibow AJ, eds. Imaging of the pancreas: cystic and rare tumors. New York, NY: Springer, 2003;97–135.

6. Procacci C, Graziani R, Bicego E, et al. Intraductal mucin-producing tumors of the pancreas: imaging findings. Radiology 1996;198(1):249–257.

7. Fukushima N, Mukai K, Sakamoto M, et al. Invasive carcinoma derived from intraductal papillary-mucinous carcinoma of the pancreas: clinicopathologic and immunohistochemical study of eight cases. Virchows Arch 2001; 439(1):6–13.

8. Jyotheeswaran S, Zotalis G, Penmetsa P, Le-

vea CM, Schoeniger LO, Shah AN. A newly recognized entity: intraductal "oncocytic" papillary neoplasm of the pancreas. Am J Gastroenterol 1998;93(12):2539–2543.

9. Albert J, Schilling D, Breer H, Jungius KP, Riemann JF, Adamek HE. Mucinous cystadenomas and intraductal papillary mucinous tumors of the pancreas in magnetic resonance cholangiopancreatography. Endoscopy 2000;32(6):472–476.

10. Procacci C, Graziani R, Bicego E, et al. Serous cystadenoma of the pancreas: report of 30 cases with emphasis on the imaging findings. J Comput Assist Tomogr 1997;21(3):373–382.

11. Dunnick NR. Image interpretation session: 1999. Intraductal mucin-producing tumor of the pancreas. RadioGraphics 2000;20(1):258–260.

12. de Lima JE Jr, Javitt MC, Mathur SC. Mucinous cystic neoplasm of the pancreas. RadioGraphics 1999;19(3):807–811.

13. Bluemke DA, Soyer P. CT of endocrine and cystic tumors of the pancreas. In: Terrier F, Grossholz M, Becker CD, eds. Spiral CT of the abdomen. New York, NY: Springer, 2000;215–226.

14. Jacobs J. Cystic pancreatic neoplasms. Acta Radiol Port 2002;53(14):35–36.

15. Stanley RJ, Semelka RC. Pancreas. In: Lee JKT, Sagel SS, Stanley RJ, Heiken JP, eds. Computed body tomography with MRI correlation. 3rd ed. Philadelphia, Pa: Lippincott-Raven, 1998;907–913.

16. Compagno J, Oertel JE. Microcystic adenomas of the pancreas (glycogen-rich cystadenomas): a clinicopathologic study of 34 cases. Am J Clin Pathol 1978;69(3):289–298.

17. Stanley RJ, Semelka RC. Pancreas. In: Lee JKT, Sagel SS, Stanley RJ, Heiken JP, eds. Computed body tomography with MRI correlation. 3rd ed. Philadelphia, Pa: Lippincott-Raven, 1998;913–914.

18. Zamboni G, Capelli P, Pesci A, Brighenti A. Pathology of cystic tumors. In: Imaging of the pancreas cystic and rare tumors. New York, NY: Springer, 2003;9–25.

19. Stanley RJ, Semelka RC. Pancreas. In: Lee JKT, Sagel SS, Stanley RJ, Heiken JP, eds. Computed body tomography with MRI correlation. 3rd ed. Philadelphia, Pa: Lippincott-Raven, 1998;902–906.

Congratulations to the five individuals who submitted the most likely diagnosis (intraductal papillary mucinous tumor of the pancreas) for Diagnosis Please, Case 136. The names and locations of the individuals, as submitted, are as follows:

Albert J. Alter, MD, PhD, *Madison, Wis*
Fahad Azzumeea, MBBS, *Montreal, Quebec, Canada*
Antonio A. Cavalcanti, MD, *Sao Paulo, Brazil*
Michael H. Childress, MD, *Silver Spring, Md*
Seyed A. Emamian, MD, PhD, *Rockville, Md*

# Radiology 2008

## This is your reprint order form or pro forma invoice

**(Please keep a copy of this document for your records.)**

Reprint order forms and purchase orders or prepayments must be received 72 hours after receipt of form either by mail or by fax at 410-820-9765. It is the policy of Cadmus Reprints to issue one invoice per order. **Please print clearly**.

Author Name _____
Title of Article _____
Issue of Journal_____ Reprint # _____ Publication Date _____
Number of Pages_____ KB # _____ Symbol Radiology
Color in Article? Yes / No (Please Circle)

**Please include the journal name and reprint number or manuscript number on your purchase order or other correspondence.**

## Order and Shipping Information

### Reprint Costs **(Please see page 2 of 2 for reprint costs/fees.)**

_____ Number of reprints ordered $_____
_____ Number of color reprints ordered $_____
_____ Number of covers ordered $_____
**Subtotal** $_____
Taxes $_____

(*Add appropriate sales tax for Virginia, Maryland, Pennsylvania, and the District of Columbia or Canadian GST to the reprints if your order is to be shipped to these locations*.)

First address included, add $32 for each additional shipping address $_____

**TOTAL** $_____

### Shipping Address **(cannot ship to a P.O. Box) Please Print Clearly**

Name _____
Institution _____
Street _____
City _____ State _____ Zip _____
Country _____
Quantity_____ Fax _____
Phone: Day _____ Evening _____
E-mail Address _____

### Additional Shipping Address* **(cannot ship to a P.O. Box)**

Name _____
Institution _____
Street _____
City _____ State _____ Zip _____
Country _____
Quantity _____ Fax _____
Phone: Day _____ Evening _____
E-mail Address _____
** Add $32 for each additional shipping address*

### Payment and Credit Card Details

**Enclosed**: Personal Check _____
Credit Card Payment Details _____
Checks must be paid in U.S. dollars and drawn on a U.S. Bank.

Credit Card: __ VISA __ Am. Exp. __ MasterCard
Card Number _____
Expiration Date_____
Signature: _____

Please send your order form and prepayment made payable to:

**Cadmus Reprints**
**P.O. Box 751903**
**Charlotte, NC 28275-1903**

***Note: Do not send express packages to this location, PO Box.***
*FEIN #:541274108*

### Invoice or Credit Card Information

**Invoice Address Please Print Clearly**
**Please complete Invoice address as it appears on credit card statement**

Name _____
Institution _____
Department _____
Street _____
City _____ State _____ Zip _____
Country _____
Phone _____ Fax _____
E-mail Address _____

**Cadmus will process credit cards and *Cadmus Journal Services* will appear on the credit card statement.**

***If you don't mail your order form, you may fax it to 410-820-9765 with your credit card information.***

Signature _____ Date _____
Signature is required. By signing this form, the author agrees to accept the responsibility for the payment of reprints and/or all charges described in this document.

RB-9/26/07

# Radiology 2008

## Black and White Reprint Prices

| Domestic (USA only) | | | | | | |
|---|---|---|---|---|---|---|
| # of Pages | 50 | 100 | 200 | 300 | 400 | 500 |
| 1-4 | $221 | $233 | $268 | $285 | $303 | $323 |
| 5-8 | $355 | $382 | $432 | $466 | $510 | $544 |
| 9-12 | $466 | $513 | $595 | $652 | $714 | $775 |
| 13-16 | $576 | $640 | $749 | $830 | $912 | $995 |
| 17-20 | $694 | $775 | $906 | $1,017 | $1,117 | $1,220 |
| 21-24 | $809 | $906 | $1,071 | $1,200 | $1,321 | $1,471 |
| 25-28 | $928 | $1,041 | $1,242 | $1,390 | $1,544 | $1,688 |
| 29-32 | $1,042 | $1,178 | $1,403 | $1,568 | $1,751 | $1,924 |
| Covers | $97 | $118 | $215 | $323 | $442 | $555 |

| International (includes Canada and Mexico) | | | | | | |
|---|---|---|---|---|---|---|
| # of Pages | 50 | 100 | 200 | 300 | 400 | 500 |
| 1-4 | $272 | $283 | $340 | $397 | $446 | $506 |
| 5-8 | $428 | $455 | $576 | $675 | $784 | $884 |
| 9-12 | $580 | $626 | $805 | $964 | $1,115 | $1,278 |
| 13-16 | $724 | $786 | $1,023 | $1,232 | $1,445 | $1,652 |
| 17-20 | $878 | $958 | $1,246 | $1,520 | $1,774 | $2,030 |
| 21-24 | $1,022 | $1,119 | $1,474 | $1,795 | $2,108 | $2,426 |
| 25-28 | $1,176 | $1,291 | $1,700 | $2,070 | $2,450 | $2,813 |
| 29-32 | $1,316 | $1,452 | $1,936 | $2,355 | $2,784 | $3,209 |
| Covers | $156 | $176 | $335 | $525 | $716 | $905 |

## Color Reprint Prices

| Domestic (USA only) | | | | | | |
|---|---|---|---|---|---|---|
| # of Pages | 50 | 100 | 200 | 300 | 400 | 500 |
| 1-4 | $223 | $239 | $352 | $473 | $597 | $719 |
| 5-8 | $349 | $401 | $601 | $849 | $1,099 | $1,349 |
| 9-12 | $486 | $517 | $852 | $1,232 | $1,609 | $1,992 |
| 13-16 | $615 | $651 | $1,105 | $1,609 | $2,117 | $2,624 |
| 17-20 | $759 | $787 | $1,357 | $1,997 | $2,626 | $3,260 |
| 21-24 | $897 | $924 | $1,611 | $2,376 | $3,135 | $3,905 |
| 25-28 | $1,033 | $1,071 | $1,873 | $2,757 | $3,650 | $4,536 |
| 29-32 | $1,175 | $1,208 | $2,122 | $3,138 | $4,162 | $5,180 |
| Covers | $97 | $118 | $215 | $323 | $442 | $555 |

| International (includes Canada and Mexico)) | | | | | | |
|---|---|---|---|---|---|---|
| # of Pages | 50 | 100 | 200 | 300 | 400 | 500 |
| 1-4 | $278 | $290 | $424 | $586 | $741 | $904 |
| 5-8 | $429 | $472 | $746 | $1,058 | $1,374 | $1,690 |
| 9-12 | $604 | $629 | $1,061 | $1,545 | $2,011 | $2,494 |
| 13-16 | $766 | $797 | $1,378 | $2,013 | $2,647 | $3,280 |
| 17-20 | $945 | $972 | $1,698 | $2,499 | $3,282 | $4,069 |
| 21-24 | $1,110 | $1,139 | $2,015 | $2,970 | $3,921 | $4,873 |
| 25-28 | $1,290 | $1,321 | $2,333 | $3,437 | $4,556 | $5,661 |
| 29-32 | $1,455 | $1,482 | $2,652 | $3,924 | $5,193 | $6,462 |
| Covers | $156 | $176 | $335 | $525 | $716 | $905 |

Minimum order is 50 copies. For orders larger than 500 copies, please consult Cadmus Reprints at 800-407-9190.

### Reprint Cover

Cover prices are listed above. The cover will include the publication title, article title, and author name in black.

### Shipping

Shipping costs are included in the reprint prices. Domestic orders are shipped via UPS Ground service. Foreign orders are shipped via a proof of delivery air service.

### Multiple Shipments

Orders can be shipped to more than one location. Please be aware that it will cost $32 for each additional location.

### Delivery

Your order will be shipped within 2 weeks of the journal print date. Allow extra time for delivery.

### Tax Due

Residents of Virginia, Maryland, Pennsylvania, and the District of Columbia are required to add the appropriate sales tax to each reprint order. For orders shipped to Canada, please add 7% Canadian GST unless exemption is claimed.

### Ordering

Reprint order forms and purchase order or prepayment is required to process your order. Please reference journal name and reprint number or manuscript number on any correspondence. You may use the reverse side of this form as a proforma invoice. Please return your order form and prepayment to:

**Cadmus Reprints**
P.O. Box 751903
Charlotte, NC 28275-1903

***Note: Do not send express packages to this location, PO Box.***
*FEIN #:541274108*

**Please direct all inquiries to:**

***Rose A. Baynard***
800-407-9190 (toll free number)
410-819-3966 (direct number)
410-820-9765 (FAX number)
baynardr@cadmus.com (e-mail)

**Reprint Order Forms and purchase order or prepayments must be received 72 hours after receipt of form.**