Pseudomyxoma peritonei (PMP) syndrome is a rare condition characterized by large amounts of mucinous ascites that accumulate in the abdomen and pelvis. Historically, the term “jelly belly” has been used to describe the gelatinous-like fluid that implants on the peritoneal surfaces and omentum (Harshen, Jyothirmayi, & Mithal, 2003; Hinson & Ambrose, 1998) (see Figure 1). PMP syndrome is considered to be a borderline malignancy because the tumor usually is not aggressive and metastasis to solid organs via the lymphatic system or bloodstream does not occur. PMP syndrome remains a fatal condition. Eventually, space in the abdomen and pelvis becomes filled with a tumor of varying consistency that causes intestinal obstruction (Hinson & Ambrose; Sugarbaker, Fernandez-Trigo, & Shamsa, 1996; Witham, 2003).

Originally, PMP syndrome was used to describe mucinous ascites associated with a ruptured appendix mucocoele. Many oncologists and pathologists have applied the term to any disease in the abdomen and pelvis characterized by mucinous fluid (Sugarbaker et al., 1997). As a result, prognosis of the disease has been variable and unpredictable.

PMP syndrome has been histologically classified into three categories: disseminated peritoneal adenomucinosis (DPAM), peritoneal mucinous carcinomatosis (PMCA), and PMCA with intermediate or discordant features (PMCA-I/D). Classification is based on the morphologic features of the epithelium identified in peritoneal lesions and the nature of the underlying lesion. DPAM describes lesions that have little cytologic atypia or mitotic activity, whereas cytologic features of carcinoma can be found in PMCA and PMCA-I/D, which are more aggressive. The classification system is important because survival rates have been found to differ significantly for each category and may affect treatment choices (Ronnett, Zahn, et al., 1995).

At a Glance

✦ Pseudomyxoma peritonei (PMP) syndrome is an uncommon, slowly progressive disease characterized by large amounts of mucinous ascites, which accumulate in the abdomen and pelvis. Five-year survival rates for PMP syndrome range from 75%–86%.

✦ Treatment options for PMP syndrome are multimodal and include observation, aggressive debulking surgery, intraperitoneal chemotherapy, radiotherapy, and mucolytic agents.

✦ Treating PMP syndrome with intraperitoneal chemotherapy allows higher concentrations of cytotoxic agents to be administered directly into abdominal and pelvic surfaces where the tumor is located without producing toxic systemic levels.

PMP syndrome should be applied only to histologically benign peritoneal tumors that are slow growing and associated most commonly with appendiceal mucinous adenoma. Pathologically, mucinous lesions are classified as DPAM or adenomucinosis (Sugarbaker et al., 1997). This article will focus on the DPAM pathologic classification.

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Pathophysiology

In PMP syndrome, an adenoma develops in the appendiceal lumen, and as the adenoma grows, the lumen of the appendix closes. Mucus normally produced by the appendiceal epithelium, as well as adenoma, causes the blockage, which results in distention of the distal portion of the appendix. As the distention increases, the wall of the appendix eventually bursts and leaks mucus, producing epithelial cells in the peritoneal cavity (Sugarbaker et al., 1997). The adenoma usually grows very slowly; however, the epithelial cells become dispersed widely throughout the peritoneal space. The cells continue to multiply and produce large amounts of mucus as the appendix repeatedly decompresses by bursting and rescaling (Esquivel & Sugarbaker, 2000). After appendiceal rupture from an adenoma, patients may present with symptoms of appendicitis.

The usual flow of peritoneal fluid moves tumor cells to the abdominal surfaces that absorb peritoneal fluid, such as the greater omentum, lesser omentum, and right hemidiaphragm. Surgical exploration has revealed that the intestinal surfaces usually have minimal mucinous seeding. Peristaltic activity of the small bowel is believed to prevent the mucinous tumor from implanting in a process referred to as the redistribution phenomena (Esquivel & Sugarbaker, 2000; Sugarbaker, 1996).

Incidence

One of every one million individuals will be diagnosed with PMP syndrome, with approximately 300 new cases diagnosed annually in the United States (Sugarbaker et al., 1997). PMP syndrome occurs two to three times more often in women than men, with a median age at diagnosis of 53 years (Hinson & Ambrose, 1998; Sherer, Abuafia, & Eliakim, 2001). PMP syndrome is found incidentally during 2 of every 10,000 laparotomies (Hinson & Ambrose).

Clinical Presentation

Symptoms vary and are dependant on the location of the disease. The most common symptom reported by men and women is a gradual increase in abdominal girth (see Figure 2). Patients complain that they are unable to lose weight even with diet and exercise and their abdomens continue to expand. The second most common symptom for men is an enlarging hernia. For women, the second most common symptom is an ovarian mass, which can be bilateral or unilateral and often is detected during a routine pelvic examination or when women present with infertility. When ovarian mucinous tumors are found, the possibility of a primary appendiceal tumor must be considered and the appendix always should be removed and microscopically examined (Galani, Marx, Steer, Culora, & Harper, 2003; Hinson & Ambrose, 1998; Ronnett, Kurman, et al., 1995; Sugarbaker, 1996).

Other symptoms may include abdominal and pelvic pain, nausea, vomiting, fatigue, changes in bowel function, and urinary tract symptoms (Galani et al., 2003; Hinson & Ambrose, 1998; Ronnett, Kurman, et al., 1995). As abdominal girth and weight increase, patients often report a decrease in nutritional intake because of pressure on the gut and a feeling of fullness, which prevent them from eating normally. Those gastrointestinal symptoms have caused some patients to be diagnosed with irritable bowel syndrome for years prior to the diagnosis of PMP syndrome.

Diagnosis

Preoperative imaging is useful and provides information necessary for planning the debulking operation. Because PMP syndrome is rare, patients should seek a surgeon who has the time, facilities, and ability to perform the radical debulking that may be necessary. In addition, the surgeon should be able to initiate adjuvant intraperitoneal (IP) chemotherapy, if recommended. Imaging also is important when evaluating for disease recurrence. Plain radiography and contrast studies are not helpful in diagnosing PMP syndrome but may be of assistance in determining bowel involvement and obstruction, especially in cases of recurrence (Harshen et al., 2003; Hinson & Ambrose, 1998; Walensky, Venbrux, Prescott, & Osterman, 1996).

Ultrasonography often is used diagnostically because it can reveal nonmobile echogenic ascites and homogeneous tumor deposits. Scalloping of hepatic and splenic margins also may be seen (Galani et al., 2003; Harshen et al., 2003; Seshul & Coulam, 1981; Walensky et al., 1996; Yeh et al., 1984). When performing diagnostic and therapeutic paracentesis, an ultrasound may be helpful because mucin often is thick and difficult to aspirate (Galani et al.). Computed tomography (CT) of the pelvis and abdomen is used most often to diagnose and determine the extent of PMP syndrome. Common findings include large amounts of mucinous material of similar density to water, which displaces the small bowel and mesenteric fat. Other findings highly indicative of PMP syndrome include thickening of the omentum, mucin septations, scalloping of hepatic and splenic margins, and curvilinear calcifications (Hinson & Ambrose, 1998; Matsumi et al., 1999; Seshul & Coulam; Sugarbaker et al., 1997; Walensky et al.) (see Figure 3). Magnetic resonance imaging (MRI) provides information similar to that obtained with a CT scan and images that help distinguish whether the fluid is mucin or ascites. Currently, no data exist to suggest that MRI is superior to CT scanning.
in diagnosing PMP syndrome. The use of MRI is limited by its cost (Buy, Malbec, Ghossain, Guinet, & Ecoiffier, 1989; Galani et al.; Harshen et al.; Walensky et al.). Cytology has not proven to be clinically valuable in diagnosing PMP syndrome. Aspirates usually contain mucus and few cells. Frozen section analysis has been used for histologic assessment of biopsies obtained during a debulking operation. Microscopically, the appearance is very similar to that of endometriosis; thus, frozen section analysis is not very helpful (Hinson & Ambrose; Pfister & Richartz, 1995). The significance of tumor markers in PMP syndrome is under investigation. Elevated tumor markers are believed to be indicative of advanced and invasive disease, which would make complete cytoreduction difficult. Moran and Cecil (2003) suggested that the standard practice should include carcinoembryonic antigen (CEA), carbohydrate antigen (CA) 19–9 and CA-125 measurements. Elevated CEA and CA 19–9 levels decreased after aggressive cytoreduction and hyperthermic IP chemotherapy. During follow-up, elevated CA 19–9 levels have proven to be more indicative of a possible recurrence (van Ruth, Hart, Bonfrer, Verwaal, & Zoetmulder, 2002). CA-125, a common ovarian cancer tumor marker in women, can be elevated in men and women with advanced metastatic intestinal adenocarcinoma (Moran & Cecil).

**Treatment**

No accepted standard treatment for PMP syndrome exists, but in most situations, multimodal therapy is delivered. Options include observation, surgery, chemotherapy, radiotherapy, and mucolytic agents. When developing a treatment plan, the extent of the disease, histology, and overall condition of the patient are considered (Galani et al., 2003).

**Observation**

Because the course of PMP syndrome often is indolent and distant metastases are extremely rare, Friedland, Allardice, and Wyatt (1986) suggested that no treatment is necessary and patients should be observed and treated symptomatically. More recent studies favor aggressive surgical intervention along with IP chemotherapy (Bryant et al., 2004; Esquivel & Sugarbaker, 2000; Gough et al., 1994).

**Surgery**

The traditional surgical approach for PMP syndrome has been debulking to remove as much of the tumor as possible. Debulking usually includes removal of the appendix, the right colon and omentum, and, in women, the ovaries and uterus (Witham, 2003). The intent is to resect all gross disease to reduce the build-up of mucus (Bryant et al., 2004). Because residual disease often remains in the peritoneal cavity, disease progression is likely. Debulking surgery can be repeated, but with each attempt, the procedure becomes more difficult because of adhesions and small bowel involvement. Tumor removal from the small bowel is difficult and can lead to serious complications, such as fistulae and malabsorption, particularly when a large portion of the small bowel is resected (Bryant et al.; Witham, 2003). Patients may survive for several years with repeated debulking procedures, but eventually no further surgical options are available and patients die from intestinal obstruction and cachexia (Esquivel & Sugarbaker, 2000; Zoetmulder & Sugarbaker, 1996).

A more aggressive approach developed by Sugarbaker, Kern, and Lack (1987) is cytoreductive surgery followed by IP chemotherapy. Cytoreductive surgery is the aggressive removal of all visible tumor deposits throughout the peritoneal cavity. IP chemotherapy may be given intraoperatively or in the early postoperative period to eliminate any microscopic residual disease that remains in the abdominal cavity. In addition, some patients receive systemic chemotherapy. The goal of the combined treatment is curative (Esquivel & Sugarbaker, 2000; Sugarbaker et al., 1987).

Cytoreductive surgery involves a series of peritonectomy procedures that remove the parietal peritoneal disease and resect some abdominal organs. Six different procedures have been described and are used depending on the distribution and volume of the tumor found at laparotomy.

- Greater omentectomy and splenectomy
- Stripping of the left hemidiaphragm
- Stripping of the right hemidiaphragm, including removal of the tumor from the liver
- Lesser omentectomy and cholecystectomy
- Antrectomy
- Pelvic peritonectomy
The six surgical procedures rarely are needed for one patient if a low volume of disease is present. Tenckhoff catheters can be placed into the abdominal cavity through the abdominal wall during surgery for the administration of IP chemotherapy either intraoperatively or postoperatively (Sugarbaker, 1995, 2006).

Cytoreductive surgery varies in duration from 5–20 hours, with an average of 10–12 hours (Deraco et al., 2004; Witham, 2003). The procedure is extensive, involving the dissection of multiple areas of the abdomen. The goal of cytoreduction is to leave no tumors greater than 2.5 mm so that cytotoxic agents are able to penetrate remaining tumor deposits (Bryant et al., 2004).

Patients must be in reasonably good health to undergo cytoreductive surgery. Patients with extensive comorbidities, such as heart and pulmonary disease, may have too many risk factors to be candidates for the operation. Radical surgical treatment is contraindicated for patients with large tumor involvement of the small bowel because they will not be left with enough bowel for adequate nutrient absorption following radical cytoreduction (Witham, 2004). Complications from this aggressive surgery are listed in Figure 4.

Morbidity rates for cytoreductive surgery range from 27%–39% and mortality rates from 2.7%–9% have been reported. Higher morbidity and mortality rates also have been associated with repeated debulking surgery attempts (Sugarbaker, 1995; Witkamp et al., 2001).

**Chemotherapy**

The treatment methodology related to chemotherapy for PMP syndrome is controversial. Treatment can include systemic chemotherapy, IP chemotherapy, and IP hyperthermic chemotherapy.

Palliative systemic chemotherapy may help with symptom control at the advanced stage of the disease when surgical options have been exhausted. Anecdotal reports have described

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**Figure 4. Surgical Complications**

<table>
<thead>
<tr>
<th>Stomach or bowel perforation</th>
<th>Intra-abdominal or wound abscess</th>
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<tbody>
<tr>
<td>Enteral fistula</td>
<td>Intestinal obstruction</td>
</tr>
<tr>
<td>Pulmonary embolism</td>
<td>Anastomotic leak</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>Postoperative bleeding</td>
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<tr>
<td>Pancreatitis</td>
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</tbody>
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**Figure 3. Abdominal and Pelvic Computed Tomography Scans**

*Note. Images courtesy of Mayo Clinic. Used with permission.*
the use of single-agent systemic chemotherapy with 5-fluorouracil, cyclophosphamide, doxorubicin, hexamethylmelamine, or cisplatin (Harshen et al., 2003).

IP administration of chemotherapy has many pharmacokinetic and toxicity advantages in comparison to systemic chemotherapy. For example, direct IP exposure to cytotoxic drugs increases the local drug potency with less systemic toxicity than IV administration. Administering chemotherapy regionally can kill any microscopic residual disease present after all of the detectable tumor has been excised. Commonly used agents for IP administration are mitomycin C, 5-fluorouracil, cyclophosphamide, paclitaxel, carboplatin, and cisplatin (Harshen et al., 2003; Sugarbaker, Mora, Carmignani, Stuart, & Yoo, 2005). Generally given in a 2,000 ml infusion, 5-fluorouracil is administered at 1,000 ml per hour for two hours via a Tenckhoff catheter. Some patients may have difficulty tolerating the amount of fluid. The efficacy of decreasing the volume is unknown. During postoperative administration of IP chemotherapy, patients remain in bed and their position is changed every 15 minutes (see Figure 5). The rotation helps ensure adequate exposure of all peritoneal surfaces at risk for residual tumor cells.

If adequate cytoreduction has been achieved, IP hyperthermic chemotherapy can be given intraoperatively. The chemotherapy agents produce a cytotoxic effect for a brief period of time and increase tumor kill with higher temperatures. Hyperthermia also can improve the diffusion of chemotherapeutic drugs into the tumor. Patients are cooled to a core temperature of 34°C–35°C and chemotherapy is heated to 40°C–43°C while perfused through catheters placed in the abdominal cavity. Typically, a flow rate of approximately 800 ml per minute is used for approximately two hours of drug exposure (Stephens, White, Esquivel, Stuart, & Sugarbaker, 2005). Agents used for IP hyperthermic chemotherapy include mitomycin C, 5-fluorouracil, oxaliplatin, etoposide, melphalan, and cisplatin (Sugarbaker et al., 2005). The benefit of the agents is difficult to assess because randomized data related to their use is limited. Based on study evidence, Sugarbaker (2006) recommended that cytoreductive surgery combined with perioperative hyperthermic IP mitomycin C followed by five days of postoperative IP 5-fluorouracil be considered the standard of care for PMP syndrome.

**Radiotherapy**

The use of external beam radiotherapy in the treatment of PMP syndrome is limited and accompanied by significant morbidity (el Sayed, 1990; Gough et al., 1994; Long, Spratt, & Dowling, 1980). A retrospective study at Mayo Clinic found an increased disease-free survival time for patients with symptomatic PMP syndrome who received IP radioisotopes after debulking surgery and IP chemotherapy, but the experience was limited and the data uncontrolled (Gough et al.). Radioisotopes used for IP therapy postoperatively include radioactive phosphorus-32, gold-198, and iodine-131.

**Mucolytic Agents**

Peritoneal lavage with a mucolytic agent, such as 10% dextrose in water, has been suggested as a way to loosen and decrease
the reaccumulation of mucus deposits and assist with the closed catheter drainage of mucin for palliation (Galani et al., 2003; Green, Gancedo, Smith, & Bernett, 1975; Harshen et al., 2003; Hinson & Ambrose, 1998). Roy, Thomas, and Horowitz (1997) reported a patient with potentially fatal hyperglycemia after the use of dextrose peritoneal lavage. The benefit of using mucolytic agents has not been confirmed by clinical studies.

### Nursing Care

Postoperatively, the care of patients who undergo cytoreductive surgery should be similar to that of patients undergoing major abdominal surgery. Appropriate antiembolism prophylaxis measures should be taken as patients are at risk for deep venous thrombosis, which may result in pulmonary embolus. Hemoglobin and
hematocrit values should be monitored along with any signs of bleeding from incision or drain sites. Patients should be observed for signs of infection as they are at greater risk for peritonitis as a result of abdominal surgery and administration of regional chemotherapy. If IP chemotherapy is administered, abdominal drain site care should be performed as needed; drain sites tend to leak. Appropriate cytotoxic drug precaution protocols should be followed to manage drainage and bodily fluids. For nursing management of adverse effects of IP chemotherapy, refer to Table 1.

Psychosocial support is important because PMP syndrome is rare, and educational resources are lacking (Witham, 2003). Prior to initiation of therapy, patients’ knowledge of the disease and treatment should be assessed. Education should include information about the surgical procedure and postoperative care, the process for administering IP chemotherapy and systemic chemotherapy, potential side effects, and the need for appropriate follow-up care (Hydzik, 2007).

Follow-Up

Patients diagnosed with PMP syndrome normally experience a prolonged clinical course. After aggressive debulking surgery and treatment, patients are monitored for disease recurrence. Follow-up includes a history and physical examination because some symptoms are signs of disease progression (Gough et al., 1994). If CA 19–9, CA-25, or CEA were elevated prior to treatment, laboratory values should be monitored every three months because an increase may indicate disease progression or recurrence (Lam et al., 2003; van Ruth et al., 2002). Follow-up CT scanning can be considered and may guide decisions regarding further surgical intervention (Levitz, Sugarbaker, Lichtman, & Brun, 2004; Smeenk, Verwaal, Antonini, & Zoetmuler, 2007; Walensky et al., 1996).

Recurrence and Survival Rates

After initial treatment, PMP syndrome recurs in most patients. Miner et al. (2005) reported a recurrence rate of 91%, of which 50% occurred within two years of initial surgery. Long-term survival is dependent on histologic grading, the completeness of cytoreductive surgery, and previous surgical interventions. For patients with complete cytoreduction, DPAM by pathology, and intraoperative or postoperative IP chemotherapy, the five-year survival rate is 75%–86%, with a 10-year survival rate of 68% (Ronnett, Yan, Kurman, Shmookler, & Sugarbaker, 2001; Sugarbaker, 2001). In 2006, Yan et al. reported a DPAM five-year survival rate of 100%.

Palliative Care

Even with aggressive treatment, the condition of many patients will deteriorate over time, requiring the focus of care to become palliative. As abdominal distension increases, progressive symptoms of obstruction and decreased nutritional intake become common. Small or more frequent meals of low-fiber foods may help decrease discomfort and complaints of early satiety. Intermittent or long-term parenteral alimentation may be used to rest the bowel and provide adequate nutrition. Nausea and vomiting can be treated with antiemetics (Mann, Wagner, Chumas, & Chalas, 1990; Witham, 2004). Patients and families will continue to require emotional support as end-of-life issues arise.

Conclusion

PMP syndrome is a rare disease that often is fatal because of the accumulation of mucinous ascites in the abdominal cavity. Although several treatment options are available, debulking surgery and the administration of IP chemotherapy have become standard treatment. Survival rates are dependent on histologic grading and the ability to aggressively remove all tumor deposits in the peritoneal cavity. Improved survival rates have been reported with cytoreductive surgery followed by IP chemotherapy, shifting the intent of treatment from palliation to potential cure. During follow-up care, patients who require further debulking operations are identified. Psychological support remains necessary because many patients eventually die from the disease.

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