Anesthetic Implications for Patients With Swyer-James Syndrome

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Objective: **The aim of this study was to describe the anesthetic management and perioperative outcomes in patients with Swyer-James syndrome (SJS), a rare acquired pulmonary disorder caused by postinfectious bronchiolitis obliterans resulting in airway obstruction and focal areas of emphysema or bronchiectasis.**

Design: **A retrospective computerized search of the medical records database at a large academic tertiary referral center was performed for patients with SJS from January 1, 2001 through July 31, 2012 who underwent procedures requiring anesthesia. A review of the perioperative course in the SJS patients as well as in those identified by a literature search was performed.**

Setting: **Academic tertiary referral center, both inpatient and outpatient settings.**

Participants: **Patients with a diagnosis of SJS.**

Interventions: **No interventions were performed.**

Measurements and Main Results: **The authors identified 4 patients with SJS who underwent 7 uneventful anesthetics including one lung resection. Three patients were symptomatic preoperatively. The literature review identified 23 SJS patients who underwent lung resection for recurrent pulmonary symptoms. One patient developed hypoxemia during one-lung ventilation. Three patients had a prolonged hospital stay (≥10 days). All patients undergoing lung resection were young (≤42 years of age).**

Conclusion: **Though SJS may be an incidental finding, these patients may have marked symptomatology, recurrent pulmonary infections, and pneumothoraces that may require lung resection. The patients tolerated anesthesia well. In severe SJS cases, pulmonary pathology and perioperative management strategies parallel that of patients with severe obstructive pulmonary disease.**

KEY WORDS: Swyer-James syndrome, anesthesia, surgery, lung disease, MacLeod's syndrome, unilateral hyperlucent lung syndrome

SWYER-JAMES SYNDROME (SJS) is a rare acquired pulmonary disorder that develops secondary to infectious etiologies in early childhood.[1] The condition manifests as a postinfectious bronchiolitis obliterans characterized with obstruction and emphysematous changes or bronchiectasis.[2–7] These changes may lead to fibrosis in both the bronchioles and interalveolar septum, resulting in pulmonary capillary bed destruction.[5,8] Clinical symptomology ranges from none in asymptomatic individuals with incidentally diagnostic findings on routine chest radiograph to severe symptoms, including dyspnea, recurrent pulmonary infections, hemoptysis, and pulmonary hypertension.[4,9] The pulmonary changes associated with this disorder could pose challenges for anesthetic management. Because this is a very rare disorder, large-scale prospective studies are not feasible. To further assess the anesthesia outcomes in patients with SJS, the authors used the medical records database of a large academic referral center to identify SJS patients who underwent anesthesia, and then conducted a literature search of patients with SJS to review their perioperative course.

METHODS

After Institutional Review Board approval, a computerized search of the medical records database at a large academic tertiary referral center from January 1, 2001 through July 31, 2012 was conducted to identify patients with SJS who underwent anesthesia. The terms Swyer-James, unilateral emphysema, emphysema unilateral, hyperlucent lung, unilateral hyperlucent lung, and hyperlucent lobe were used to identify patients with this condition. Only patients with a confirmed diagnosis of SJS by a pulmonologist were included in the investigation. Patient records were reviewed for demographic variables, preoperative pulmonary status and symptoms, and other comorbid conditions. The surgical and anesthetic records were reviewed for details regarding the perioperative course. Hospital records were reviewed for postoperative events within 30 days with emphasis on respiratory complications.

To review the perioperative complications in patients with SJS, the authors performed a literature search using an OVID-based search strategy of (1) PubMed (2001-present), (2) MEDLINE (2001-present), (3) EMBASE (2001-present), and (4) Web of Science (2001-present) under the following keywords: Swyer-James syndrome, Swyer-James-MacLeod, unilateral hyperlucent lung syndrome, all limited to humans and English language. Bibliographies of identified published reports were searched for additional references. Identified reports of patients undergoing surgery were reviewed to identify perioperative complications.

RESULTS

In the medical records database, 14 patients with SJS were identified, and all had provided authorization for search of their medical records.[10] Of these 14 patients, 4 underwent a total of 7 anesthetics at this institution and will be referred to as group I (Table 1). Three of four patients had respiratory symptoms preoperatively. One patient had dyspnea on exertion, another had exertional dyspnea and hemoptysis, and a third patient had hemoptysis and recurrent pneumonias. Three of the procedures performed on these patients were related to the diagnosis of SJS. There were no major perioperative complications related to SJS. Postoperative complications were related to the procedure and were not related to pulmonary pathology.

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Table 1. Patient Descriptions and Perioperative Findings in Patients with SJS Undergoing Anesthesia from the Medical Records Database at This Tertiary Referral Center

Patient #/Sex	Patient 1 Female	Patient 2 Female	Patient 3 Male	Patient 4 Female
Procedure/ Age	1. [a] Craniotomy 36 yo 2. Cesarean section 40 yo	1. [a] Bronchoscopy 69 yo 2. Radius osteotomy 56 yo	1. [a] Cardiac catheterization (radiofrequency ablation for atrial fibrillation) 41 yo 2. Bronchoscopy 46 yo	Right upper and middle lobe lobectomy 22 yo
Comorbidities	1. Meningioma, Seizures 2. None	1. None 2. None	1. Atrial fibrillation 2. Fibrosing mediastinitis 3. Severe pulmonary hypertension	Recurrent pneumonia Previous smoker
Preoperative Symptoms	1. None 2. None	1. Dyspnea 2. Dyspnea	1. Palpitations, dyspnea related to atrial fibrillation 2. Hemoptysis, dyspnea	Hemoptysis
Perioperative Complications	1. Dilantin toxicity 2. None	1. None 2. None	1. Pericardial effusion requiring pericardiocentesis 2. None	None Resected lung grew *Mycobacterium avium* complex
Discharge POD	1. POD 5 2. POD 3	1. POD 0 2. POD 2	1. POD 5 2. POD 0	POD 8
Anesthesia Type	1. General anesthesia 2. Epidural anesthesia	1. MAC[b] 2. General anesthesia	1. General anesthesia 2. MAC[b]	General anesthesia
Premedication	1. Midazolam 2. None	1. Midazolam 2. None	1. Midazolam 2. Midazolam	Midazolam
Induction Medications	1. Thiopental 2. None	1. None 2. Thiopental	1. Thiopental 2. None	Propofol
Muscle Relaxant	1. Succinylcholine, pancuronium 2. N/A	1. None 2. Succinylcholine	1. None 2. None	Succinylcholine Vecuronium
Intraoperative Analgesic	1. Fentanyl infusion 2. Lumbar epidural	1. Topical lidocaine 2% Fentanyl 2. Oxymorphone	1. Fentanyl 2. Fentanyl	Thoracic epidural
Maintenance Anesthetic	1. Nitrous oxide/fentanyl infusion 2. Epidural anesthesia	1. N/A 2. Isoflurane	1. Isoflurane/nitrous oxide 2. Propofol	Isoflurane
Antiemetics	1. Droperidol 2. Droperidol	1. None 2. Droperidol	1. Ondansetron 2. Ondansetron	Ondansetron
Airway Management	1. OTT 2. 10 L Oxygen via close face mask	3. 1.OTT* 4. 2.OTT	1. OTT 2. OTT*	Double-lumen OTT
Ventilator Mode	1. AC 2. N/A	1. Spontaneous respirations 2. AC	1. AC 2. Spontaneous respirations	AC
Tidal Volume	1. 9mL/kg 2. N/A	1. N/A 2. 10 mL/kg	1. NR 2. N/A	6 mL/kg
Peek Pressure	1. 32 cmH_20 2. N/A	1. N/A 2. N/A	1. NR 2. N/A	22-28 cmH_20 during OLV
F_iO_2	1. 0.30 2. N/A	1. 1.0 2. 0.40-0.60	1. 0.30 2. 1.0	1.0 during OLV
PEEP	1. None 2. N/A	1. N/A 2. 5 cmH_20	1. 5 cmH_20 2. N/A	5 cm H_20
Intraoperative Hypoxemia ($SaO_2 < 88\%$)	1. None 2. None	1. None 2. None	1. None 2. None	None
Monitors	1. Standard ASA, RAL 2. Standard ASA	1. Standard ASA 2. Standard ASA, RAL, BIS	1. Standard ASA, RAL, BIS 2. Standard ASA	Standard ASA, RAL
Extubation Location	1. OR 2. N/A	1. OR 2. OR	1. OR 2. OR	PACU
Postoperative Analgesia	1. Codeine/ acetaminophen acetaminophen 2. Propoxyphene/ acetaminophen	1. None 2. Morphine PCA codeine/ acetaminophen	1. Fentanyl in PACU 2. None	Thoracic epidural Hydromorphone PCA Oxycodone

Abbreviations: AC, assist control; ASA, American Society of Anesthesiologists; BIS, bispectral index; MAC, monitored anesthesia care; N/A, not applicable; NR, not recorded; OLV, one-lung ventilation; OTT, Oral Tracheal Tube; PACU, postanesthesia care unit; PCA, patient controlled analgesia; POD, postoperative day; RAL, radial arterial line; yo, year old.

[a]1, 2 refer to first and second operation

[b]MAC for bronchoscopy includes fiberoptic endotracheal intubation via bronchoscope after airway topicalization.