male breast, such as fibromas and
leiomyomas, are much less common but
should also be considered in the differen-
tial diagnosis.

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Correspondence: Dr. D. Aranovich, Dept. of
General Surgery A, Rabin Medical Center
(Bellinson Campus), Petah Tiqva 49202, Israel.
Phone: (972-3) 937-6210
Fax: (972-3) 937-7486
email: fgaef@daltot.org.il

Persistent Plastic Bronchitis in a Child after Cardiac Surgery

Uri Peleg MD1, Shepard Schwartz MD2, Gisela Sirota MD2, Ilan Hochman MD1, David Cohen MD1
and Elie Picard MD3,4

Departments of 1Otolaryngology and Head and Neck Surgery and 2Pediatrics, and 3Division of Pediatric Pulmonology, Shaare Zedek Medical
Center, Jerusalem, Israel
Affiliated to Faculty of Health Sciences, Ben-Gurion University of the Negev, Beer Sheva, Israel

Key words: plastic bronchitis, children, bronchoscopy, Fontan procedure

Plastic bronchitis is a rare clinical entity
that occurs as a complication of asthma,
allergy, respiratory tract infections, cystic
fibrosis, sickle cell disease, pericarditis and
congenital heart defects [1,2]. Most cases
related to cardiac defects are reported to
have occurred after corrective surgery,
mainly the Fontan procedure [1-3]. The
Fontan procedure creates a direct connec-
tion between the systemic venous circula-
tion, most frequently the superior and
inferior vena cava, and the pulmonary
artery. Plastic bronchitis presents with
severe respiratory distress and is usually
diagnosed when white casts are expecto-
rated or detected by bronchoscopy [1]. We
describe a patient who developed fatal
plastic bronchitis after undergoing the
Fontan procedure for repair of tricuspid
atresia.

Patient Description
A 6 year old girl was born with a cyanotic
heart defect that was neither diagnosed
nor treated in her country of birth. Until the
age of 3 years she suffered from chronic
hypoxia, which was the probable cause of
her mild psychomotor retardation. At age
3, immediately after she immigrated to
Israel, she was diagnosed as suffering from
tricuspid atresia with a single ventricle.
During the next 3 years she underwent a
series of operations: a right modified
Blalock-Taussig procedure, placement of
an aortopulmonary shunt, placement of a
bidirectional Glenn shunt, and finally, the
Fontan procedure. After her last surgery
she developed pneumonia in the right
lung, atelectasis of the left lung, and
chyllothorax, all of which resolved. She also
sustained permanent paralysis of her right
diaphragm. Echocardiography revealed
normal left ventricle function, mild mitral
regurgitation, and good flow through the
Fontan conduit. She was discharged home
in good condition and treated with diure-
tics and coumadin.

Two months after being discharged and
3 months after undergoing the Fontan
procedure, she arrived at the pediatric
emergency room of our hospital with
severe dyspnea and cyanosis. According
to her parents' report she developed a
cough 2 weeks previously. She was afebrile,
her respiratory rate was 60 per minute and
blood pressure 120/80. Oxygen saturation
on room air was 90%, and 88-90% with an
oxygen mask. On auscultation, breath
sounds were reduced over the right lung
field. No clinical signs of heart failure,
significant abdominal findings or clubbing
were noted. A complete blood count
revealed a white blood cell count of
15,600 with 89% neutrophils, and hemoglo-
in and platelets within the normal
range. There was no infiltrate, atelectasis or
hyperinflation on the chest X-ray, but
the right diaphragm was elevated.

She was hospitalized in the pediatric
intensive care unit and initially treated with
oxygen, chest physiotherapy, bronchodila-
tors and intravenous antibiotics. The next
day her condition improved significantly
and she was transferred to the pediatric
ward. Four days later her respiratory state
worsened and she was returned to the
pediatric intensive care unit. Chest X-ray
showed partial atelectasis of the right lung.
Emergency flexible bronchoscopy demon-
strated a cast blocking the entrance to the
right main bronchus, which was immedi-
ately removed by rigid bronchoscopy. The cast material was solid mucous in the form of a mold of the bronchial tree (Figure). Her respiratory status later improved considerably. Cultures taken from the cast were negative. Pathologic examination of the cast revealed a protein exudate mixed with few neutrophils and lymphoid cells. During the same hospital stay there were repeated episodes of respiratory deterioration, each requiring bronchoscopic removal of similar casts from the right bronchial tree. Therapy with inhalation of acetylcysteine and urokinase, as well as with systemic steroids and azithromycin was unsuccessful in preventing further cast formation.

In an effort to improve the effectiveness of her cough and her ability to clear cast material from her right lung, plication of the right diaphragm was performed. This too failed to improve her course. Cardiac catheterization revealed slightly reduced cardiac output and normal pressures in the Fontan circulation.

In the ensuing months she was hospitalized every 2-3 weeks for similar episodes of respiratory distress, all treated by bronchoscopic removal of newly formed casts. Treatment with tissue plasminogen activator inhalation during one hospitalization showed no clear benefit. During her final hospital stay, and 7 months after the Fontan procedure, she developed severe respiratory failure with a room air oxygen saturation of 80%. Rigid bronchoscopy was performed, during which she suffered a cardiopulmonary arrest. Despite all resuscitative efforts, she died a few hours later.

Comment

To the best of our knowledge the incidence of plastic bronchitis is not mentioned in the literature, though it is considered a rare entity with only approximately 50 cases published to date [1-5]. We report here a 6 year old girl born with a congenital cyanotic heart defect, who developed plastic bronchitis 3 months after a Fontan procedure and died 4 months later. Despite treatment with various medications and multiple rigid bronchoscopies for removal of newly formed casts, she succumbed to her disease. We speculate that the large cast caused her severe hypoxia prior to the last bronchoscopy and initiated the rapid deterioration that worsened during the procedure and caused the child's death.

Seer et al. [2] in 1997 reported nine cases of plastic bronchitis and suggested a classification of two types according to the cast composition. Type I refers to inflammatory casts that contain mainly fibrin with a rich inflammatory cell infiltrate; these casts appear in inflammatory lung diseases. Classified as type II are acellular casts composed primarily of mucin, with few inflammatory cells; this type occurs in congenital heart defects after surgical repair. In pulmonary diseases, type I casts are formed by the affected bronchial airways. The mechanism accounting for the overproduction of mucin by the respiratory epithelium in type II cannot be directly explained by the pathophysiologic changes that occur before or after surgical correction of congenital heart defects. Seer and colleagues [2] suggested that the underlying cause is pulmonary venous hypertension; however, Languepin et al. [3] proposed lymphatic leakage in the chest as the cause of cast formation.

The prognosis is largely determined by the etiology. Brogan and associates [1] found that patients with a congenital cardiac defect had a mortality rate of 29% and a life-threatening events rate of 41%. No deaths or life-threatening events occurred among patients whose underlying disease was asthma or allergy [1]. Languepin et al. [3] described 14 patients who developed plastic bronchitis secondary to congenital heart defects, of whom 43% died [3]. Seer's group [2] claimed that if correction of cardiac dysfunction is impossible, then the prognosis is poor [2].

Treatment of type I disease includes empirical administration of systemic or inhaled steroids, antibiotics, bronchodilators and mucolytics. For type II disease the medical treatment is more problematic. There are a few sporadic reports in the literature of successful treatment of type II plastic bronchitis using a variety of treatment modalities. Inhalation of urokinase or tissue plasminogen activator, and intratracheal instillation of rhDNase led to improvement in some patients, though their effect on mucin is unclear [1]. Both oral azithromycin and subcutaneous heparin when administered long term proved effective in individual cases [4,5]. In our patient, all medications failed to prevent recurrence of casts.

The purpose of drug therapy is to prevent reproduction of casts. However, once formed, the casts must be removed with a rigid bronchoscope as their stiffness precludes use of a flexible one. Our patient underwent immediate rigid bronchoscopy for each respiratory exacerbation. In most instances, a single solid mucous cast in the form of a mold of the bronchial tree was removed either intact or in smaller pieces. In light of the high rate of treatment failure, for severe cases that develop in children after repair of congenital cardiac defects, cardiopulmonary transplant should be considered in the early stages.

In conclusion, we describe a young girl who developed plastic bronchitis after the Fontan procedure, and despite aggressive management she succumbed from this disease. Unfortunately, the optimal treatment for the prevention of bronchial acellular cast formation has yet to be found. It is important to recognize this potentially fatal complication of cardiac surgery for congenital defects. The pathophysiologic mechanism of this process requires further investigation so that a better prognosis may be achieved.
Cervico-Facial Emphysema and Pneumomediastinum Complicating a High-Speed Drill Dental Procedure

Sharon Tamir MD, George Backleh MD, Kassem Hamdan MD and Ron Eliashar MD
Department of Otolaryngology/Head & Neck Surgery, Hadassah Medical Center and Hebrew University School of Medicine, Jerusalem, Israel

Key words: emphysema, pneumomediastinum, dental drill

Pneumomediastinum and cervico-facial emphysema associated with a dental procedure was first reported by Turnbull in 1900 [1]. He described a case of massive cervico-facial subcutaneous emphysema and mediastinal emphysema following a tooth extraction in a bull. Physiologically, PMCFE can occur as a complication of coughing, sneezing, nose-blowing and vomiting, or following a dental extraction [2,3]. PMCFE occurring during dental procedures while using high-speed air or water drills has been documented mainly in the dentistry literature. The use of a high-speed drill can cause entrance of air that dissects through the periodontal ligament or under the raised mucoperiosteal flap. This air enters the sublingual and sub mandibular spaces; these spaces are in communication with the pterygomandibular, parapharyngeal and retropharyngeal spaces. Entrance of air into the gums can thus penetrate through the cervical facial planes and into the mediastinum [3].

We present a patient suffering from PMCFE with a computed tomography scan of the neck and mediastinum. As seen on the CT scan, the organs in the anterior half of the neck are surrounded by air that had dissected into the parapharyngeal region and into the mediastinum.

Patient Description
A 79 year old healthy woman underwent a surgical extraction of her right mandibular second molar at a dental clinic. During the treatment, in which a high-speed, air turbine drill was employed, the patient complained of a swelling in the neck and difficulty breathing, but suffered no chest pain. She was transferred to the emergency room.

Upon arrival her vital signs were normal. A clinical examination revealed a previously sutured right intraoral sublingual laceration. Gross facial swelling with right periorbital tissue fluctuation was noted, accompanied by cervical subcutaneous emphysema. Both lungs were clear to auscultation and no cardiac murmur or rub was detected. The rest of the physical examination was unremarkable. The electrocardiogram was normal. A chest X-ray demonstrated pneumomediastinum and subcutaneous emphysema. No pneumothorax was detected.

A contrast-enhanced axial CT scan of the neck and chest demonstrated a significant amount of free air in the facial spaces of the neck (Figure A) and in the mediastinum (Figure B). PMCFE was diagnosed. The patient was admitted to the Department of Otolaryngology/Head and Neck Surgery for airway monitoring and parenteral antibiotic therapy. She was treated with intravenous amoxicillin-clavulanic acid 1 g 3 times a day and analgesics as required. The swelling and the subcutaneous emphysema subsided within 2 days. During this period the patient did not experience any airway obstruction. She was discharged from the hospital 48 hours later and continued the oral antibiotic therapy for 7 more days with no further complications.

Comment
Symptoms of pneumomediastinum and subcutaneous emphysema in the cervico-facial region can vary. In the medical literature we find descriptions of a variety of symptoms such as swelling, a sensation of fullness in the facial/ cervical region, erythema, crepitation, dysphagia, dysphonia, dyspnea, periorbital swelling, emphysema around the eye, and pain [2–5].

The differential diagnosis of a rapid facial swelling following a dental procedure should take into consideration angioedema, hematoma, cellulitis, allergic reaction...