



国家儿童医学中心
National Center for Children's Health, China



北京儿童医院
BEIJING CHILDREN'S HOSPITAL

MY STORY IN CHIBA

-Mini-fellowship funded by APPS

Hao Wang

Respiratory Medicine Department

National Center for Children's Health

Capital Medical University Beijing Children's Hospital

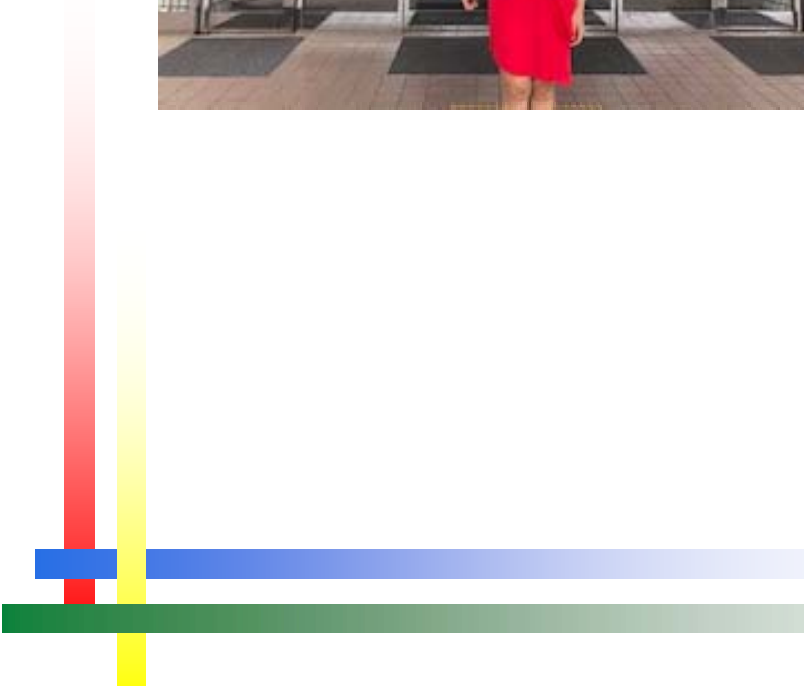


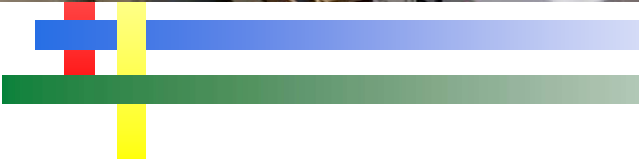
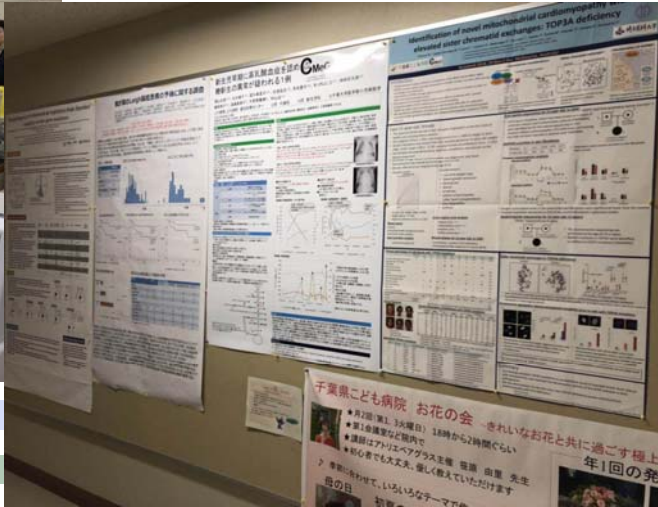
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CIPP XVIII





Nation

Specific Diagnosis Possible

Bronchiolitis Obliterans **NEHI** **Alveolar Growth Disorder**

Treatable Traits

Levent 2018; 352: 866-79

Infections

- *P. aeruginosa*
- *H. influenzae*
- *Biofilms*

Underlying disease

- Immunodeficiency
- Aspiration
- Structural airway lesions

Inflammation and airway secretions

- Eosinophilic, neutrophilic
- Airway clearance
- Mucolytics

Co-morbidities

- Airflow obstruction
- Extra-pulmonary (e.g. sleep-related disorders, upper airway disease)

Generic modifiable factors

- Hygiene practices
- Malnutrition
- Vaccinations, etc

Treatable (therapeutic) traits

- Chronic airway infection
- Antibiotic therapy
- Inhaled
- Targeted
- Macrolides
- Pathogen acquisition
- Pseudoimmune sensitization therapy
- Immunomodulatory
- Immunoglobulin replacement
- Prophylactic antibiotics
- NIM
- Antibiotic therapy
- ABPA
- Corticosteroids
- Immunoglobulin

Effectiveness and functional impairment

- Airway hyper-reactivity
- Airway hyper-responsiveness
- Airway hyper-sensitivity
- Airway hyper-irritability
- Airway hyper-sensitivity
- Airway hyper-irritability
- Airway hyper-sensitivity
- Airway hyper-irritability

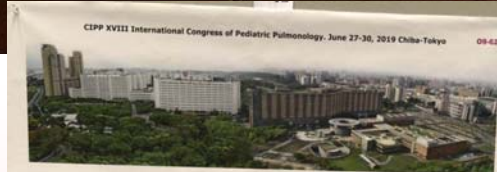
Targetable (endophenotypic) traits

- Microbial (bacterial) dysbiosis
- Probiotics
- Mycobacterium (fungal) dysbiosis
- Antifungals
- Neutrophil dysfunction
- Neutrophil elastase inhibitors
- Protease-mediated lung damage
- Protease inhibitors
- Ciliary dysfunction (primary or secondary)
- Airway clearance
- CFTR potentiator therapy
- Systemic inflammation and vascular dysfunction
- Anti-inflammatory therapy
- CFTR dysfunction
- CFTR potentiators
- CFTR correctors
- Innate immune deficiency
- TLR-based therapeutics
- Antibiotic prophylaxis

Other factors

- Ethnic differences
- Environmental exposures
- Climatic variation
- Lifestyle
- Psychosocial

James D. Chalmers, Sangy H Chotmalal
Lancet Respir Med 2018



Surfactant deficiency in a late preterm neonate: A rare presentation

Fang-Chih Lin¹, Hsiao-Ping Wang¹, Jyh-Seng Wang², Bao-Ren Nong¹

¹Department of Pediatrics, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan
²Department of Pathology, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan

INTRODUCTION

Pulmonary surfactant is essential to avert neonatal respiratory distress. It is a mixture of phospholipids and proteins which is necessary for lowering surface tension and preventing and resolving lung collapse. Surfactant protein deficiency might result from four different types of mutations of surfactant protein genes (SP-B, SP-C, SP-A, SP-D). The presentation and prognosis of these gene mutations are highly diverse. Among one of them, surfactant protein C deficiency seems a well-recognized clinical manifestation.

CASE PRESENTATION

This patient was a 36.47 weeks gestational age singleton 2700g male premature infant born to a mild chronic G6PD(A) indigenous Taiwanese mother via Caesarian. She was transferred to our neonatal care unit due to respiratory distress developed after birth. Maternal and birth history were unremarkable. Initially, nasal CPAP (continuous positive airway pressure) was provided for respiratory support, however, intubation with mechanical ventilation support was performed after one week of admission. Due to fluctuated respiratory condition and increased oxygen need, Figure 1 shows her chest X-ray film and high-resolution computed tomography in combination of her clinical presentation and image feature. Lung biopsy was performed due to diffuse lung disease was highly suspected. Figure 2 and 3 present the pathological findings. Distended and atrophic alveoli were demonstrated but the effect was not obvious. Hyaline membrane was spared because of her G6PD (glucose 6-phosphate dehydrogenase) deficiency. Parents refused the suspension of lung transplantation. Due to worsening of clinical condition and resistance to medication treatment, she received palliative intubation after full discussion with her family at 6th month. The result of next generation sequencing (NGS) and SRA sequence analysis showed heterozygous mutation of SP-C (c.1235G>A) which is a mutation on SP-C gene. Surfactant protein C deficiency was then confirmed.

DISCUSSION

There is no standard or protocol treatment for surfactant protein C deficiency. Although surfactant protein C deficiency and alveolar interstitial lung disease due to its pathophysiological effects. The drug was first indicated due to her underlying G6PD deficiency.

CONCLUSION

In conclusion, even with some supportive treatment, surfactant protein C deficiency present with diverse pulmonary phenotype and prognosis especially the initial presentation was bronchopulmonary dysplasia syndrome in premature neonates. We should raise our awareness of surfactant protein deficiency in neonates with atypical clinical course of respiratory symptoms, atypical histology finding and unresponsive to standard treatment of respiratory distress syndrome. Further study is needed to clarify the exact mechanism of gene mutation pathogenesis and accurate presentation of this disease.

Figure 1: Series of chest radiograph and HRCT. Figure 2: Histology of lung biopsy. Figure 3: Histology of lung biopsy.

Early Diagnosis of Infantile Laryngeal and Tracheal Cyst by Flexible Bronchoscopy: case reports

Ti-An Tsai, Chang-Ku Tsai, Yi-Chen Lee, Ta-Yu Liu, Chih-Min Tsai, Chen-Kuang Niu and Hong-Ren Yu

Department of Pediatrics, Chang Gung Memorial Hospital - Kaohsiung Medical Center, Taiwan

Case Presentation

Case 1: This is a 3-month-old boy who suffered from stridor breathing sounds since 1-month-old accompanied with poor feeding and failure to thrive. Bronchoscope and Head and Neck computed tomography revealed one 1.1x1.3 cm laryngeal cyst at midline and another 6mm cyst inferior to epiglottis. The patient underwent CO₂ laser marsupialization and the pathology showed mucocutaneous tissue with cyst lined by stratified squamous epithelium.

Case 2: A corrected age of 10-month-5-day-old boy had symptoms of dyspnea and stridor but no fever. Croup was the initial impression, but the treatment response was poor. Then bronchoscope and Head and Neck computed tomography were arranged. One 6x3 mm subglottic endotracheal lesion causing airway compromised was noted. He underwent CO₂ laser marsupialization and the pathology showed fragmented mucosal and fibrous tissue focally lined by respiratory-type ciliated epithelial cells.

Conclusion

Early flexible bronchoscopy is a helpful examination for diagnosis of infantile stridor. Epidermoid cyst, thyroglossal cyst, and valvular cyst should be considered in the differential diagnosis of congenital laryngeal cyst.

