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A CLINICAL GUIDELINE FOR STRUCTURED ASSESSMENT OF CT-IMAGING IN CONGENITAL LUNG ABNORMALITIES.


ABSTRACT

OBJECTIVES: To develop a clinical guideline for structured assessment and uniform reporting of congenital lung abnormalities (CLA) on Computed Tomography (CT)-scans.

MATERIALS AND METHODS: A systematic literature search was conducted for articles describing CT-scan abnormalities of congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), congenital lobar emphysema (CLE) and bronchogenic cyst (BC). A structured report using objective features of CLA was developed after consensus between a pediatric pulmonologist, radiologist and surgeon.

RESULTS: Of 1581 articles identified, 158 remained after title-abstract screening by two independent reviewers. After assessing full-texts, we included 28 retrospective cohort-studies. Air-containing cysts and soft tissue masses are described in both CPAM and BPS while anomalous arterial blood supply is only found in BPS. Perilesional low-attenuation areas, atelectasis and mediastinal shift may be found in all aforementioned abnormalities and can also be seen in CLE as a cause of a hyperinflated lobe. We have developed a structured report, subdivided into five sections: Location & Extent, Airway, Lesion, Vascularization and Surrounding tissue.

CONCLUSIONS: CT-imaging findings in CLA are broad and nomenclature is variable. Overlap is seen between and within abnormalities, possibly due to definitions often being based on pathological findings, which is an unsuitable approach for CT imaging. We propose a structured assessment of CLA using objective radiological features and uniform nomenclature to improve reporting.

SURFACTANT PROTEIN DISORDERS IN CHILDHOOD INTERSTITIAL LUNG DISEASE.


ABSTRACT

Surfactant, which was first identified in the 1920s, is pivotal to lower the surface tension in alveoli of the lungs and helps to lower the work of breathing and prevents atelectasis. Surfactant proteins, such as surfactant protein B and surfactant protein C, contribute to function and stability of surfactant film. Additionally, adenosine triphosphate binding cassette 3 and thyroid transcription factor-1 are also integral for the normal structure and functioning of pulmonary surfactant. Through the study and improved understanding of surfactant over the decades, there is increasing interest into the study of childhood interstitial lung diseases (chILD) in the context of surfactant protein disorders. Surfactant
protein deficiency syndrome (SPDS) is a group of rare diseases within the chILD group that is caused by genetic mutations of SFTPB, SFTPC, ABCA3 and TTF1 genes. Conclusion: This review article seeks to provide an overview of surfactant protein disorders in the context of chILD. What is Known: • Surfactant protein disorders are an extremely rare group of disorders caused by genetic mutations of SFTPB, SFTPC, ABCA3 and TTF1 genes. • Given its rarity, research is only beginning to unmask the pathophysiology, inheritance, spectrum of disease and its manifestations. What is New: • Diagnostic and treatment options continue to be explored and evolve in these conditions. • It is, therefore, imperative that we as paediatricians are abreast with current development in this field.

THE EFFICACY AND SAFETY OF MONTELUKAST IN CHILDREN WITH OBSTRUCTIVE SLEEP APNEA: A SYSTEMATIC REVIEW AND META-ANALYSIS.


https://doi.org/10.1016/j.sleep.2020.11.009

ABSTRACT

OBJECTIVE The efficacy and safety of montelukast in children with obstructive sleep apnea (OSA) remain controversial. Therefore, the aims of this systemic review and meta-analysis are to verify this issue and further provide reference for clinical practice.

METHODS Seven databases were searched for randomized controlled trials (RCTs) up to September 30, 2019. The literature screening and data extraction were performed by two independent researchers. Adverse reactions from trials were also recorded. Meta-analysis was performed and analyzed heterogeneity. Methodological and evidence quality were followed by to evaluate according to Cochrane handbook.

RESULTS A total of 4 RCTs including 305 children with mild to moderate OSA were involved. Compared with placebo, we found that oral montelukast (OM) significantly improved polysomnography (PSG) monitoring parameters, typical and relevant symptoms including snoring and mouth breathing, and adenoid morphology in children with OSA. When compared with routine drugs, not only PSG monitoring parameters and adenoid morphology, but also sleep-disordered breathing (SDB)-related questionnaire scores were improved in patients with OSA treated by combination of OM and routine drugs. In addition, compared with single nasal spray of mometasone furoate, the present study also showed that OM combined with nasal spray of mometasone furoate significantly improved PSG monitoring parameters, symptoms of snoring and mouth breathing and reduced tonsil morphology in pediatric OSA. In terms of treatment safety, one study reported adverse reactions of OM such as headache, nausea and vomiting, while no adverse events were reported after OM treatment in another study.

CONCLUSION As a classic leukotriene receptor antagonist, montelukast can be used to treat children with mild to moderate OSA in the short term and improve clinical characteristics. The promotion and application of OM in clinic is considered to be a noninvasive option to avoid surgical treatment.
IMPACT OF ORAL CORTICOSTEROIDS ON RESPIRATORY OUTCOMES IN ACUTE PRESCHOOL WHEEZE: A RANDOMISED CLINICAL TRIAL.


ABSTRACT

OBJECTIVE: To determine if administration of oral prednisolone to preschool children with acute wheeze alters respiratory outcomes.

DESIGN: Double-blind, randomised, placebo-controlled equivalence trial.

SETTING: Three hospitals in New Zealand.

PATIENTS: 477 children aged 24-59 months with acute wheeze associated with respiratory illness.

INTERVENTIONS: 2 mg/kg (maximum 40 mg) oral prednisolone or similar placebo, once daily for 3 days.

MAIN OUTCOME MEASURES: Primary outcome was change in Preschool Respiratory Assessment Measure (PRAM) score 24 hours after intervention. Secondary outcomes included PRAM score at 4 hours, length of emergency department and inpatient stays, admission and representation rates, time to return to normal activities and use of additional oral prednisolone or intravenous medications. Analysis was by intention-to-treat.

RESULTS: There was no difference between groups for change in PRAM score at 24 hours (difference between means -0.39, 95% CI -0.84 to 0.06, p=0.09). Absolute PRAM score was lower in the prednisolone group at 4 hours (median (IQR) 1 (0-2) vs 2 (0-3), p=0.01) and 24 hours (0 (0-1) vs 0 (0-1), p=0.01), when symptoms had resolved for most children regardless of initial treatment. Admission rate, requirement for additional oral prednisolone and use of intravenous medication were lower in the prednisolone group, although there were no differences between groups for time taken to return to normal activities or rates of representation within 7 days.

CONCLUSION: Oral prednisolone does not alter respiratory outcomes at 24 hours or beyond in preschool children presenting with acute wheeze.

THE AIRWAY MICROBIOME AND PEDIATRIC ASTHMA.


ABSTRACT
PURPOSE OF REVIEW: Asthma is the most common chronic disease of childhood. Investigations of the lower and upper airway microbiomes have significantly progressed over recent years, and their roles in pediatric asthma are becoming increasingly clear.

RECENT FINDINGS: Early studies identified the existence of upper and lower airway microbiomes, including imbalances in both associated with pediatric asthma. The infant airway microbiome may offer predictive value for the development of asthma in later childhood, and it may also be influenced by external factors such as respiratory viral illness. The airway microbiome has also been associated with the clinical course of asthma, including rates of exacerbation and level of control. Advances in -omics sciences have enabled improved identification of the airway microbiome’s relationships with host response and function in children with asthma. Investigations are now moving toward the application of the above findings to explore risk modification and treatment options.

SUMMARY: The airway microbiome provides an intriguing window into pediatric asthma, offering insights into asthma diagnosis, clinical course, and perhaps treatment. Further investigation is needed to solidify these associations and translate research findings into clinical practice.

SPONTANEOUS PRIMARY PNEUMOMEDIASTINUM: IS IT ALWAYS BENIGN?
Alemu, B. N., E. T. Yeheyis and A. G. Tiruneh

ABSTRACT
Spontaneous Pneumomediastinum is a rare disease. It could be a simple and self-limited condition or be a life-threatening complication of underlying diseases. The therapeutic options also differ by the cause. This systematic review was done to provide, as far as we know, the first attempt to broadly assess the clinical feature, predisposing factors, possible management, and outcome of spontaneous primary pneumomediastinum.

OUTCOMES OF PROTRACTED BACTERIAL BRONCHITIS IN CHILDREN: A 5-YEAR PROSPECTIVE COHORT STUDY.

ABSTRACT
BACKGROUND AND OBJECTIVE Long-term data on children with PBB has been identified as a research priority. We describe the 5-year outcomes for children with PBB to ascertain the presence of chronic respiratory disease (bronchiectasis, recurrent PBB and asthma) and identify the risk factors for these.

METHODS Prospective cohort study was undertaken at the Queensland Children’s Hospital, Brisbane, Australia, of 166 children with PBB and 28 controls (undergoing bronchoscopy for symptoms other
than chronic wet cough). Monitoring was by monthly contact via research staff. Clinical review, spirometry and CT chest were performed as clinically indicated.

**RESULTS** A total of 194 children were included in the analysis. Median duration of follow-up was 59 months (IQR: 50–71 months) post-index PBB episode, 67.5% had ongoing symptoms and 9.6% had bronchiectasis. Significant predictors of bronchiectasis were recurrent PBB in year 1 of follow-up (ORadj = 9.6, 95% CI: 1.8–50.1) and the presence of Haemophilus influenzae in the BAL (ORadj = 5.1, 95% CI: 1.4–19.1). Clinician-diagnosed asthma at final follow-up was present in 27.1% of children with PBB. A significant BDR (FEV1 improvement >12%) was obtained in 63.5% of the children who underwent reversibility testing. Positive allergen-specific IgE (ORadj = 14.8, 95% CI: 2.2–100.8) at baseline and bronchomalacia (ORadj = 5.9, 95% CI: 1.2–29.7) were significant predictors of asthma diagnosis. Spirometry parameters were in the normal range.

**CONCLUSION** As a significant proportion of children with PBB have ongoing symptoms at 5 years, and outcomes include bronchiectasis and asthma, they should be carefully followed up clinically. Defining biomarkers, endotypes and mechanistic studies elucidating the different outcomes are now required.

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**DRUG-INDUCED SLEEP ENDOSCOPY DIRECTED SURGERY IMPROVES POLYSOMNOGRAPHY MEASURES IN OVERWEIGHT AND OBESE CHILDREN WITH OBSTRUCTIVE SLEEP APNEA.**


**ABSTRACT**

**BACKGROUND** Obstructive sleep apnea affects approximately 1–4% of all children, with increased prevalence amongst overweight and obese children.

**OBJECTIVE** To assess the effects of drug-induced sleep endoscopy (DISE)-directed surgery on polysomnography parameters in obese and overweight children.

**MATERIAL/METHODS** A retrospective case-series was performed on obese and overweight pediatric patients who underwent clinically indicated DISE-directed surgery. Forty children met the inclusion criteria, including: body mass index ≥85%, DISE-study, and pre- and post-DISE polysomnography. Patients were divided into surgically naïve (n = 23) and prior adenotonsillectomy (n = 17) groups. Demographic and clinical characteristics were examined with chi-square and Wilcoxon rank-sum test. Polysomnography parameters were compared with Wilcoxon signed rank test.

**RESULTS** Of 40 children with mean BMI 94% and mean age 8 ± 6 years old, 17 (43%) underwent a previous adenotonsillectomy. Overall, significant improvements were observed in the apnea-hypopnea index (AHI; 25.0 to 9.9 events/hour, p < .01) and oxygen nadir (82.7% to 88.5%, p < .01). A similar pattern was observed among the surgically naïve (AHI: 35.9 to 12.7 events/hour, p = .04; oxygen nadir: 79.7% to 86.4%, p = .2) and post-adenotonsillectomy groups (AHI: 10.4 to 6.2 events/hour, p = .02; oxygen nadir: 86.7% to 91.2%, p < .01).

**CONCLUSIONS/SIGNIFICANCE** Polysomnography parameters significantly improved following DISE-directed interventions in obese and overweight children with obstructive sleep apnea.
**RESPIRATORY TRAJECTORIES IN TYPE 2 AND 3 SPINAL MUSCULAR ATROPHY IN THE ISMAC COHORT STUDY.**


[https://n.neurology.org/content/96/4/e587](https://n.neurology.org/content/96/4/e587)

**ABSTRACT**

**OBJECTIVE** To describe the respiratory trajectories and their correlation with motor function in an international pediatric cohort of patients with type 2 and nonambulant type 3 spinal muscular atrophy (SMA).

**METHODS** This was an 8-year retrospective observational study of patients in the International SMA Consortium (iSMAc) natural history study. We retrieved anthropometrics, forced vital capacity (FVC) absolute, FVC percent predicted (FVC%), and noninvasive ventilation (NIV) requirement. Hammersmith Functional Motor Scale (HFMS) and revised Performance of Upper Limb (RULM) scores were correlated with respiratory function. We excluded patients in interventional clinical trials and on nusinersen commercial therapy.

**RESULTS** There were 437 patients with SMA: 348 with type 2 and 89 with nonambulant type 3. Mean age at first visit was 6.9 (±4.4) and 11.1 (±4) years. In SMA type 2, FVC%P declined by 4.2%/y from 5 to 13 years, followed by a slower decline (1.0%/y). In type 3, FVC%P declined by 6.3%/y between 8 and 13 years, followed by a slower decline (0.9%/y). Thirty-nine percent with SMA type 2% and 9% with type 3 required NIV at a median age 5.0 (1.8–16.6) and 15.1 (13.8–16.3) years. Eighty-four percent with SMA type 2% and 80% with type 3 had scoliosis; 54% and 46% required surgery, which did not significantly affect respiratory decline. FVC%P positively correlated with HFMS and RULM scores in both subtypes.

**CONCLUSIONS** In SMA type 2 and nonambulant type 3, lung function declines differently, with a common leveling after age 13 years. Lung and motor function correlated in both subtypes. Our data further define the milder SMA phenotypes and provide information to benchmark the long-term efficacy of new treatments for SMA.

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**HOME RESPIRATORY POLYGRAPHY IN OBSTRUCTIVE SLEEP APNEA SYNDROME IN CHILDREN: COMPARISON WITH A SCREENING QUESTIONNAIRE.**


**ABSTRACT**

**METHODS** This was an 8-year retrospective observational study of patients in the International SMA Consortium (iSMAc) natural history study. We retrieved anthropometrics, forced vital capacity (FVC) absolute, FVC percent predicted (FVC%), and noninvasive ventilation (NIV) requirement. Hammersmith Functional Motor Scale (HFMS) and revised Performance of Upper Limb (RULM) scores were correlated with respiratory function. We excluded patients in interventional clinical trials and on nusinersen commercial therapy.

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**CONCLUSIONS** In SMA type 2 and nonambulant type 3, lung function declines differently, with a common leveling after age 13 years. Lung and motor function correlated in both subtypes. Our data further define the milder SMA phenotypes and provide information to benchmark the long-term efficacy of new treatments for SMA.
**OBJECTIVES** The prevalence of obstructive sleep apnea syndrome (OSAS) in children referred for sleep-disordered breathing reaches up to 59%. We aimed to test the adequacy of a questionnaire compared to home respiratory polygraphy (HRP), in 45 subjects (5–16 years-old), without maxillofacial malformations nor other comorbidities, presenting with symptoms compatible with OSAS.

**METHODS** All children passed a 12-items questionnaire (Obstructive Airway Child test: OACT) and the HRP. OSAS was classified in severity according to the apnea-hypopnea index (AHI).

**RESULTS** With HRP, 60% and 15% children were detected to have at least mild (AHI ≥1) and moderate (AHI >5) OSAS, respectively. The sensitivity of the questionnaire to detect mild and moderate OSAS was good (93% and 71%, respectively) but the specificity was very low (11% and 34%). However, an OACT score under 61 showed a very good negative predictive value for moderate and severe OSAS (87%). With the questionnaire, we could have avoided a complementary PSG or HRP in 25/45 (56%) of our subjects as in children with mild OSAS and without comorbidities only clinical observation is usually advised.

**CONCLUSIONS** The OACT questionnaire has shown to be a good and quick instrument to exclude moderate and severe OSAS in our population of children without maxillofacial malformations. Indeed children scoring under 61 could avoid a constraining and expensive sleep exam. However, if the score is above this cut-off, the performance to recognize OSAS is low and the child’s evaluation must be completed by a HRP or PSG.