392 Brave lung: non invasive ventilation (NIV) and pneumothorax

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Hypercapnic respiratory failure and recurrent pneumothorax (pnx) in severe cystic fibrosis (CF) requires *ad hoc* evaluation and ventilatory management. Pnx is a relative contraindication to NIV so its use is not encouraged if pulmonary bubbles and medical history of past pnx are described.
The patient is a 26 yo male with severe lung disease (FEV_1 20–30% pred.) in nocturnal oxygen therapy (OT) at NIV start. Clinical history is characterised by malnutrition with PEG for enteral feeding, chronic airway infection by *P. aeruginosa* and MRSA, sinusopathy and depressive syndrome. In 1999 he had recurrent left pnx treated with pleurodesis and a right one ten yrs after drained and talced. A fall in lung function resulting in symptomatic hypoxaemia-hypercapnia, persistent during exacerbations, followed the last pnx. Though large bubbles all through the superior right lobe on CT Thorax, we gradually initiated him to NIV with nasal mask, IPAP and EPAP 11–4 cmH_2O in spontaneous mode. BGA motivated us to go on so we acclimatised him to night NIV with OT 1 L/min. NIV was well tolerated with disappearance of morning headache, somnolence and better quality of sleep. After a month, IPAP was set at 15 cmH_2O and awakening BGA revealed better PaO_2 and stable CO_2. After 6 months, BGA improved more: awakening CO_2 was yet ~50 mmHg and PaO_2 was fair; CT follow-up did not change. Adherence showed 100% of use for $\geqslant$4 hrs/day with an average of 9^h26^{min}.
Proven and potential benefits of NIV in CF are well known. Indications and contraindications have not been yet defined but, in our opinion, NIV can be used in severe lung damage with positive results. Nevertheless a prudent and a low-pressures-approach is mandatory.

393* CPAP for atelectasis in bronchopulmonary aspergillosis

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CPAP is a valid tool in various conditions. Physiotherapy (pt) often takes advantages by PAP devices for different purposes, such as lung recruitment manoeuvres, restrictive status and airway patency. However, there are no guidelines on how and for how long patients need to be treated with CPAP, neither clinical indications for cystic fibrosis.
A 12 years old CF girl (FEV_1 120–140% pred.) with a right lobar atelectasis started ab therapy i.v. on May 3rd 2010 with ceftazidime and tobramycin; pt program after this new finding consisted in PEP-Mask twice a day with 2.0 mm resistor and range pressure around 15–20 cmH_2O for 15 min/session, β_2 pMDI (2 puff/twice a day), DNase (1 vial/day). On May 5th i.v. therapy was changed with meropenem, levofloxacin and methylprednisolone due to increased PCR. On May 17th X-ray showed the same atelectasic area. Transferred in stable clinical conditions to our hospital, laboratory found high IgE level for *Aspergillus* suggesting bronchopulmonary aspergillosis then treated successfully with steroids and itraconazole (already in therapy). Hence we introduced then CPAP delivered by nasal mask at 10 cmH_2O for 8 hrs/day (4 hrs during afternoon and 4 hrs overnight consecutively). Patient tolerated well the CPAP treatment. After 8 days the X-ray showed a significant resolution and patient was discharged. PEP-Mask continued home in the same modality. The follow-up X-ray, two months later, showed disappearance of lung atelectasis.
CPAP therapy was determinant in accelerating this process and it resulted as a valid tool for improving lobar atelectasis. Its application is feasible in this population. Further studies are urgently needed to guide our management.

394 Cystic fibrosis liver disease: an emerging clinical issue showing a multifactorial pathogenesis

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Objectives: Cystic Fibrosis Liver Disease (CFLD) has gained clinical relevance with the decreasing mortality from extrahepatic causes. Several factors have been associated with the development of CFLD, as pancreatic insufficiency and severe genotype.
Methods: We discuss clinical and diagnostic implications of CFLD describing two case-reports. Case 1: a six-year-old male with a positive newborn screening for CF but a negative sweat test. He had pancreatic sufficiency and he was carrier of the F508del mutation at the first-level genetic analysis. He complained of recurrent episodes of bronchitis, treated with several antibiotic courses. An unexpected liver involvement (moderate steatosis, regenerative nodules) was occasionally detected at the age of 4 years. We performed the extensive genetic analysis that identified a second CFTR mutation (R31E), supporting a CF diagnosis. Case 2: a five-year-old female having constantly normal sweat test values and positive CFTR gene analysis (G542X/F1074R). She had pancreatic sufficiency but she was treated with enteral nutrition because of a failure to thrive. A liver involvement was identified since the first month of life because she had recurrent hypertransaminasemia. For this reason, she underwent a liver biopsy that showed periductal damage and biliary stasis.
Conclusions: CFLD may start early in life of CF patients. It can also occur in patients with pancreatic sufficiency. The clinical expression may be influenced by non-genetic factors, as malnutrition and antibiotic courses. A possible role of genetic modifiers is suggested by the detection of a normal sweat test. Their identification could allow prophylactic strategies.