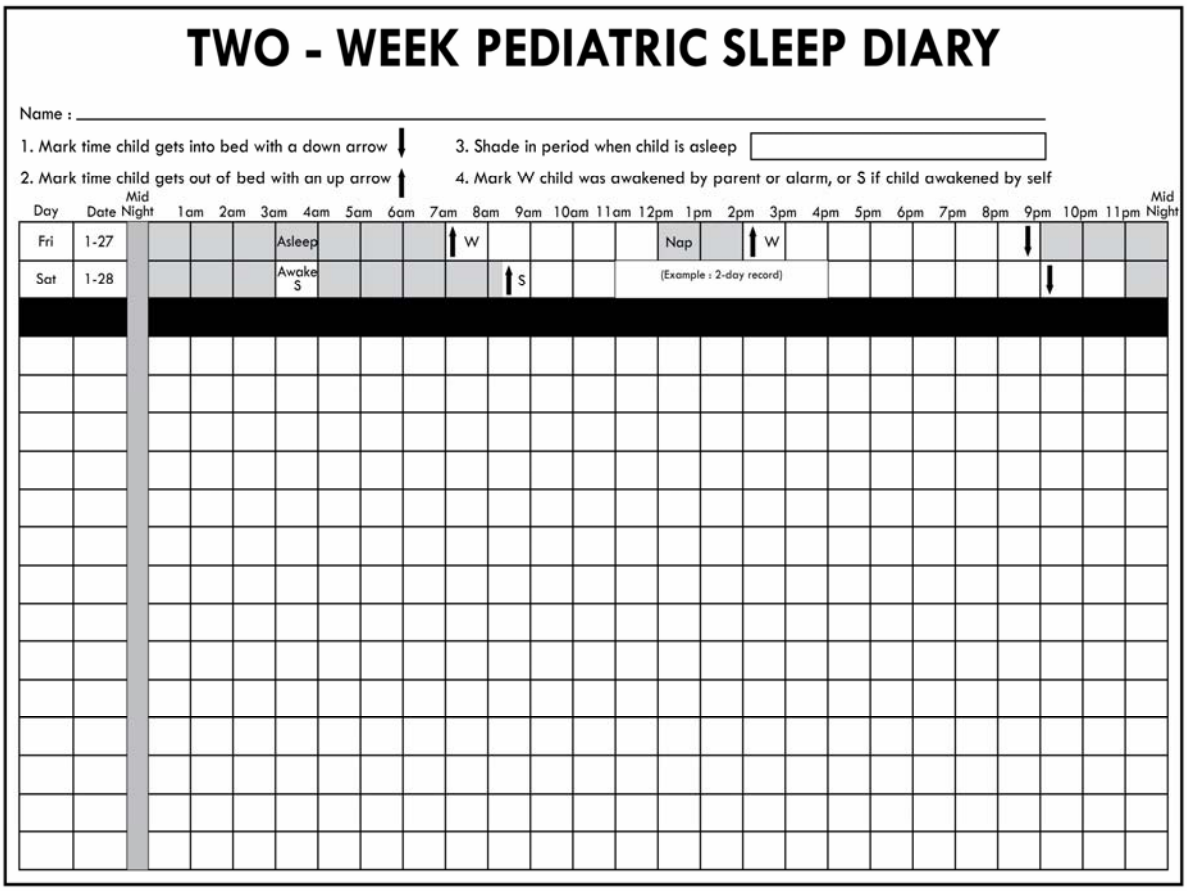
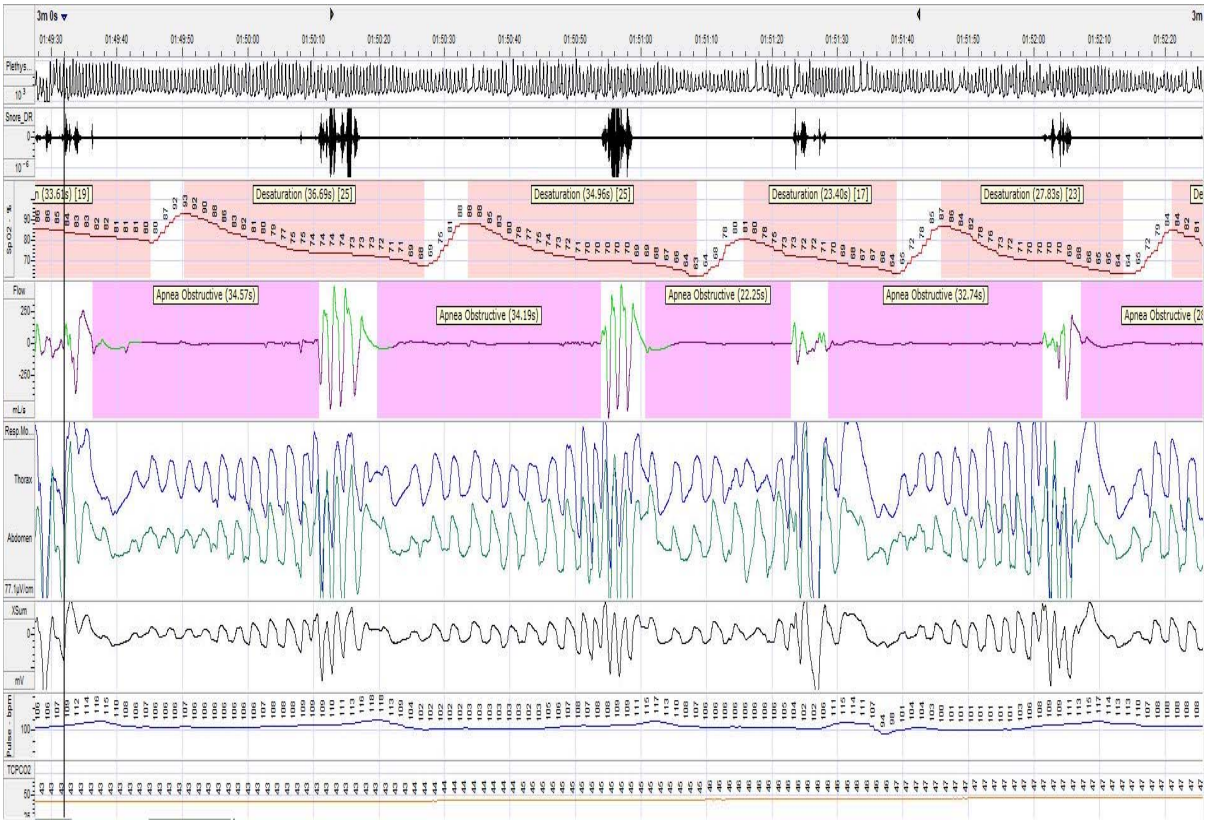
Web Table I Complex Sleep Disordered Breathing in Children

Condition	*Factors predisposing to SDB*	*Recommendation for testing*
Craniosynostosis and Craniofacial syndromes (Apert, Crouzon, Pfeiffer, Muenke and Saethre Chotzen)	Midface hypoplasia. Chiari malformation is a frequent finding in Crouzon (70%) & Pfeiffer (50%) syndromes and can have associated central sleep apnea	Yearly screening in patient with Apert, Crouzon and Pfeiffer syndromes. Muenke and Saethre Chotzen syndromes evaluate only if they become symptomatic
Down's syndrome	Relative macroglossia, midface hypoplasia, reduced muscle tone, small upper airway, adeno-tonsillar hypertrophy, obesity	All children before age of 4 years. In all children pre adenotonsillectomy
Cleft lip/palate	Small size of pharyngeal airways & craniofacial dimensions. Oropharyngeal musculature is disrupted by the cleft, which impacts on speech and swallow, as well as adversely affects maintenance of airway patency during sleep.	Sleep study is recommended if child is symptomatic, in the presence of syndromic cleft lip/palate or pre pharyngoplasty and pharyngeal flap surgery.
Syndromic micrognathia (i.e. Pierre Robin Sequence, Stickler syndrome, etc)	Micrognathia, glossoptosis, midface hypoplasia	All children should be screened
Obesity syndromes (eg Prader Willi, Bardet Biedl)	Obesity, hypotonia, micrognathia, small naso and oropharynx, scoliosis leading to OSA. Central sleep apneas, nocturnal hypoventilation, hypersomnolence/cataplexy also seen	Yearly sleep studies are recommended. Sleep study pre growth hormone therapy & post 3-6 m post therapy & afterwards if symptoms of SDB reappear. MSLT if hypersomnolence
Mucopolysaccharidosis	Cranial and spinal abnormalities (e.g., flattened nasal bridge, short neck, mandibular abnormalities) and glycosaminoglycans deposition in the mouth, nose, throat	All patients should be evaluated at diagnosis
Achondroplasia	OSA: Facial hypoplasia, retruded position of chin, adeno-tonsillar hypertrophy. Central sleep apneas also seen.	Should be screened at least once from 1 yr and then subsequently (or prior) if symptoms develop
Duchenne muscular dystrophy, limb-girdle muscular dystrophy, fascio-scapulo-humeral	Decreased respiratory drive, muscle weakness; pharyngeal muscle weakness, scoliosis, obesity, recurrent lung infections. OSA, central apneas & hypoventilation seen	FVC <60% predicted, symptoms of nocturnal hypoventilation or when children become non-ambulatory. Follow up annually.
Spinal Muscular Atrophy Congenital Myopathies Congenital Muscular Dystrophies Myotonic Dystrophy Congenital myasthenic syndromes Hereditary Motor Sensory Neuropathy	Same as above. OSA, central apneas & hypoventilation seen	Sleep study advised if: profound weakness with non ambulation, weak cry, ineffective cough, swallowing difficulties, repeated chest infections, poor lung function, scoliosis & chest wall deformity Episodic apnoea of infancy and childhood (sometimes life-threatening) described in CHAT and RAPSN mutations. Progressive respiratory muscle weakness in COLQ and DOK7 mutations. Sleep Study recommended if these mutations present and symptoms suggestive of SDB. Required if weakness is severe and persistent or if symptoms suggestive of SDB

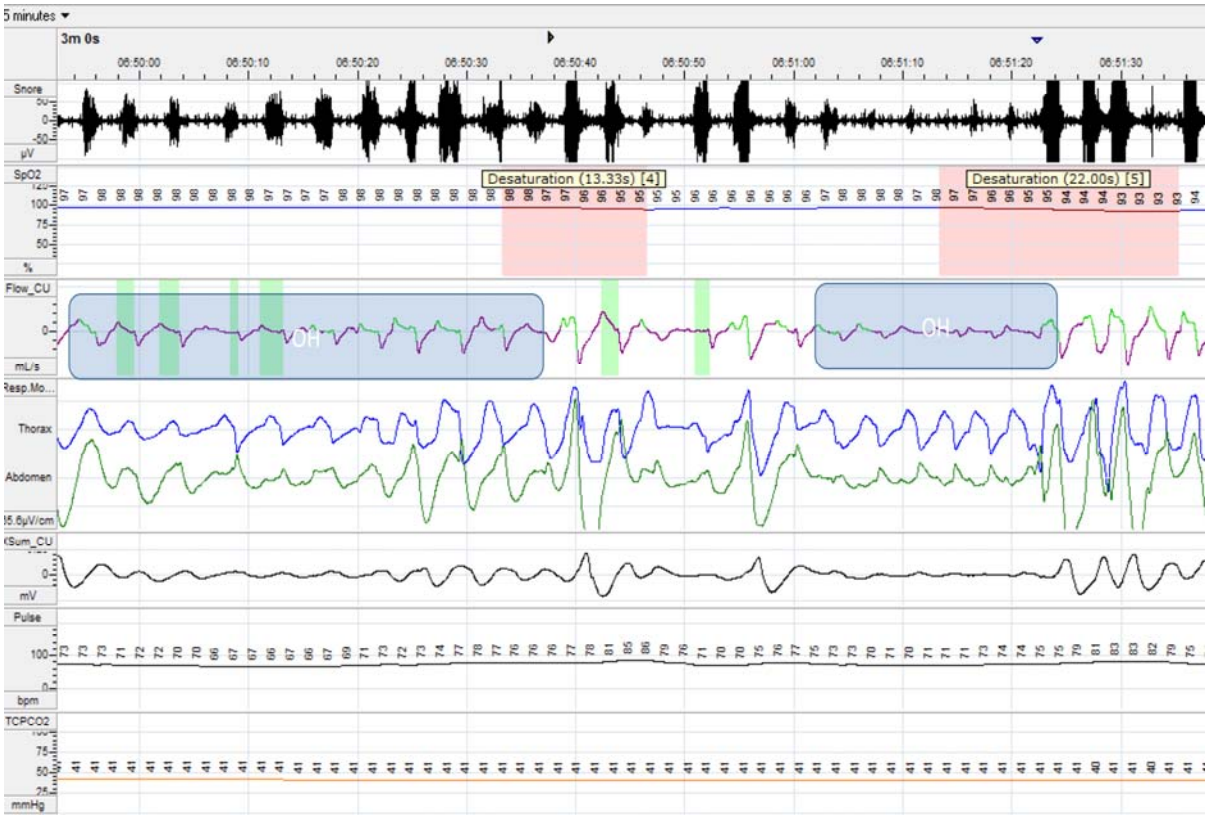


Record of sleep and naps in 24 hours; A record of 2 weeks is useful; Time at which child goes to bed is marked as ↓; Time at which child gets out of bed as ↑; Time period during which child is asleep is shaded; W signifies child is awake, either by parents or alarm or S if awakened by self.

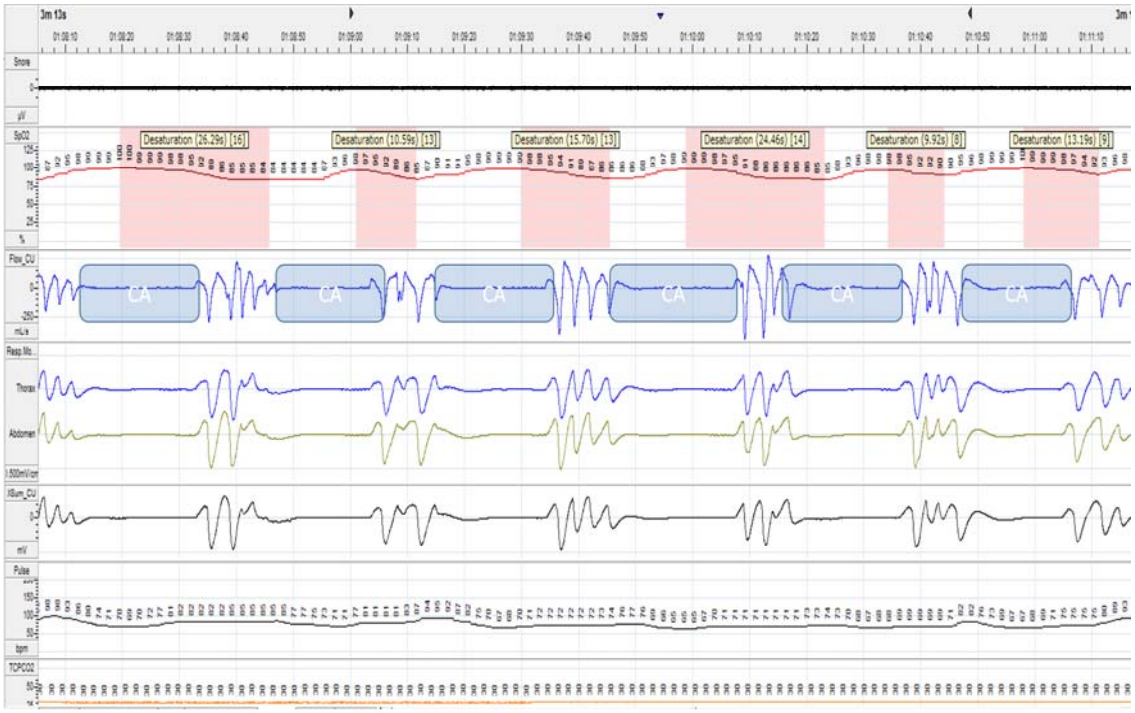
Web Fig. 1 Sleep Diary



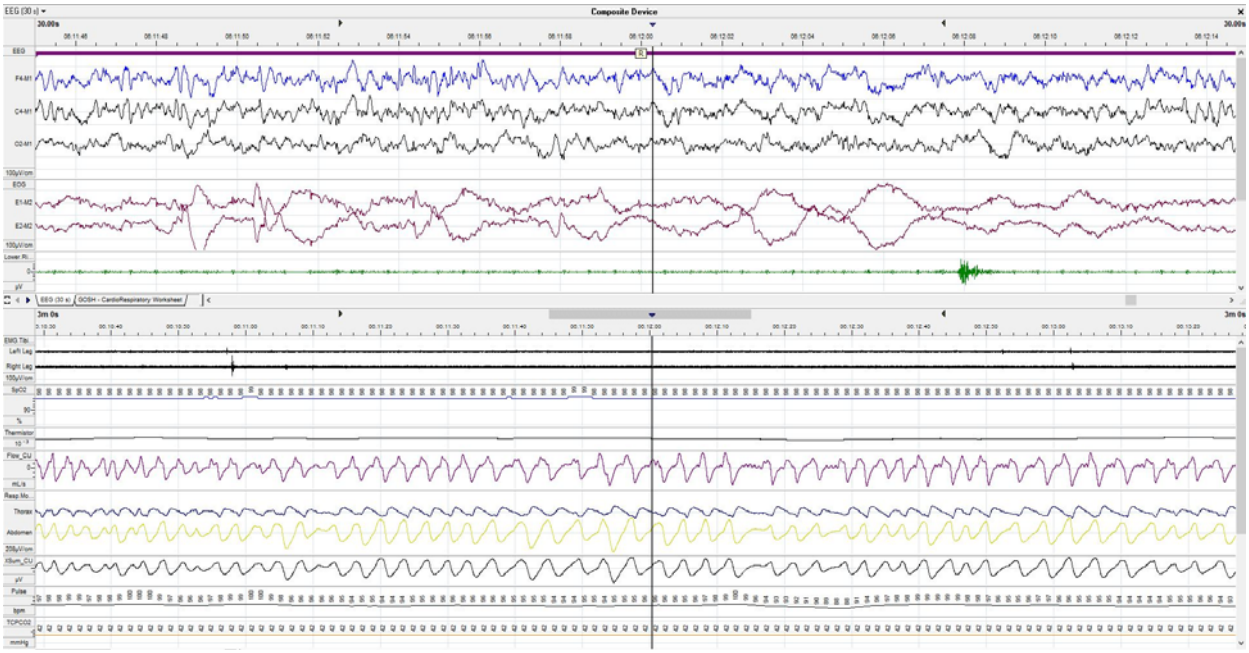
Web Fig. 2 A 3 minute epoch of a Cardiorespiratory Sleep study showing multiple Obstructive Apneas (scored on the nasal flow trace). Note the accompanying desaturations and paradoxical breathing in the thoraco-abdominal bands.



Web Fig. 3 A 3 minute epoch of a Cardiorespiratory Sleep study showing obstructive Hypopnea. Note that the flow is reduced >30% associated with desaturation and the thoraco-abdominal bands show paradoxical efforts.



Web Fig. 4 A 3 minute epoch of a Cardiorespiratory Sleep study showing multiple Central Apneas. Note that the thoraco-abdominal bands show no efforts compared to paradoxical efforts in obstructive apneas.



Web Fig. 5 A 30 s epoch of a Polysomnography. Note the additional neurological channels (EEG, EOG and chin EMG)