3rd INTERNATIONAL PEDIATRIC
NONINVASIVE VENTILATION CONFERENCE

Necker university hospital
Paris - France
November 7th & 8th 2019

Complex OSA

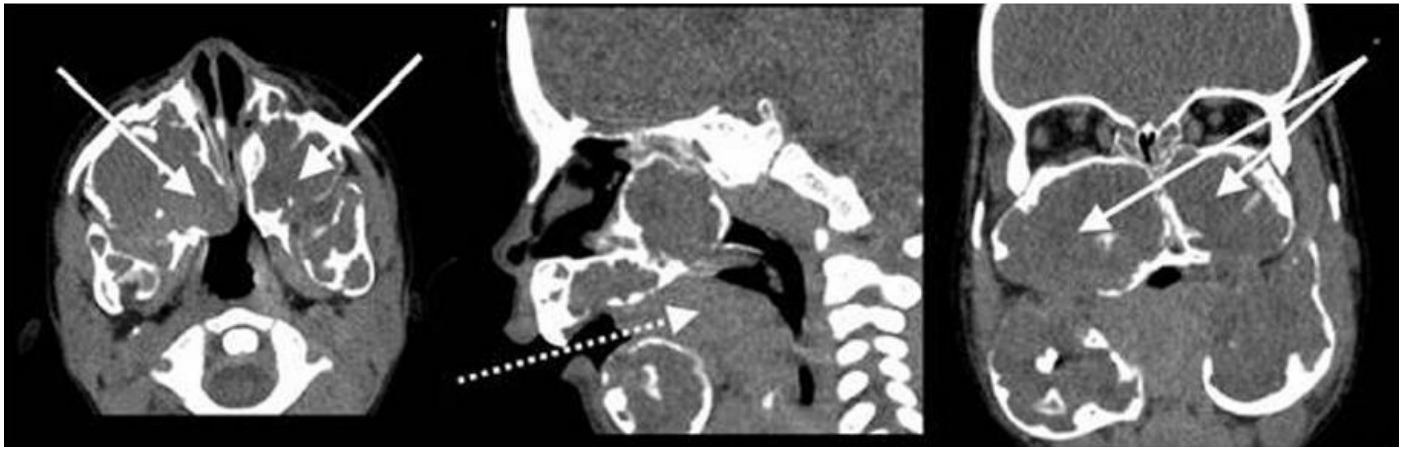
Pr Brigitte Fauroux and Pr François Abel

Université de Paris
Necker ENFANTS MALADES HÔPITAL UNIVERSITAIRE
IRBA
antadir INDÉPENDANCE ET EXPERTISE POUR LE PROGRÈS DE LA SANTÉ À DOMICILE

Case 1

4-yr old boy with cherubinism

- Cherubinism: rare hereditary disorder characterized by progressive, painless, bilateral enlargement of the mandible and/or maxilla resulting from replacement of bone with multilocular granuloma, causing firm protuberant intra-alveolar masses and teeth displacement



Sleep study

- Obstructive AHI 20 events/h
- Mean SpO_2 95%
- Oxygen desaturation index 31 events/h
- 5% of nighttime with SpO_2<90%

➔ indication of CPAP treatment (no surgical options)

Which interface would you chose ?

- Nasal mask ?
- Nasobuccal mask ?
- Mouthpiece ?
- Full face mask ?
- Nasal prongs ?

None !

- The patient could not be approached and was terrified by the idea to have anything on his face
- What would you propose ?

Nocturnal Mouthpiece Ventilation and Medical Hypnosis to Treat Severe Obstructive Sleep Apnea in a Child With Cherubism

Sonia Khirani, PhD,[1] Natacha Kadlub, MD,[2,3,4] Vincent Delord, MSN,[5] Arnaud Picard, MD, PhD,[2,3,4] and Brigitte Fauroux, MD, PhD[1,5]*

Adult mouthpiece interface (Easy Resp, Air Liquide Medical Systems, Antony, France)

CHEST

Original Research

SLEEP DISORDERS

Medical Hypnosis as a Tool to Acclimatize Children to Noninvasive Positive Pressure Ventilation

A Pilot Study

Vincent Delord, MSN; Sonia Khirani, PhD; Adriana Ramirez, MSc; Erick Louis Joseph, AS; Clotilde Gambier, MSN; Maryse Belson, MSN; Francis Gajan, MD; and Brigitte Fauroux, MD, PhD

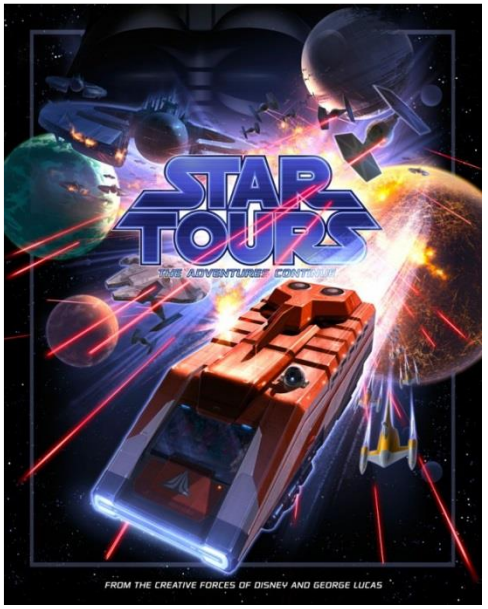
Infants (Aged About < 2-3 y)	Older Children (Aged > 2-3 y)
Distraction	Indirect suggestions
Visual stimulation	The child expresses his or her imagination spontaneously, and the hypnosis practitioner accompanies the child in the imaginary experience.
Auditory stimulation	
Kinesthetic stimulation	
	Direct suggestions
	The child is unable to express his or her imagination spontaneously, so the hypnosis practitioner gives the child direct suggestions to help him or her enter an imaginary experience.

Patients

Pat.	Age-gender	Pathology	History	Imaginary experience	Sessions	Compliance
1	7-boy	Apert syndrome	7 neurosurgical and maxillofacial interventions	space tour in a fighter	3	7
2	4-boy	Cherubinism	repeated mandibular surgery with tumour enucleations, orbital decompression, and nasal airway calibration	tour in the pirate boat with Peter Pan followed by a space tour in a rocket	3	6
3	13-boy	Franceschetti syndrome	tracheotomy since birth	floating in the sea	3	7
4	8-boy	Laryngeal paralysis	tracheotomy since birth	surfing in the sea while listening to his favorite music	3 (+1)	8
5	2-boy	Bronchopulmonary dysplasia	severe lung disease, repeated hospitalizations	game with soap bubbles and songs with the mother	1	12
6	13-girl	Cystic fibrosis	end-stage lung disease with severe dyspnea	tour in a shopping center	3	4
7	15-boy	Morbid obesity with OSAS	severe depression	attendance of a rock concert	1	-
8	5-boy	Obesity with OSAS	none	attendance of his favorite amusement park	1	10
9	6-girl	OSAS	none	attendance of a princess ball	3	8.5

7-yr old boy with Apert syndrome

7 neurosurgical and maxillofacial interventions



8-yr old Greek boy with laryngeal paralysis and tracheotomy since birth

5-yr old boy with OSA and obesity

Nasobuccal masks

Amara View (Philips)

Dreamwear (Philips)

AirFit F 30 (Resmed)

Total face masks

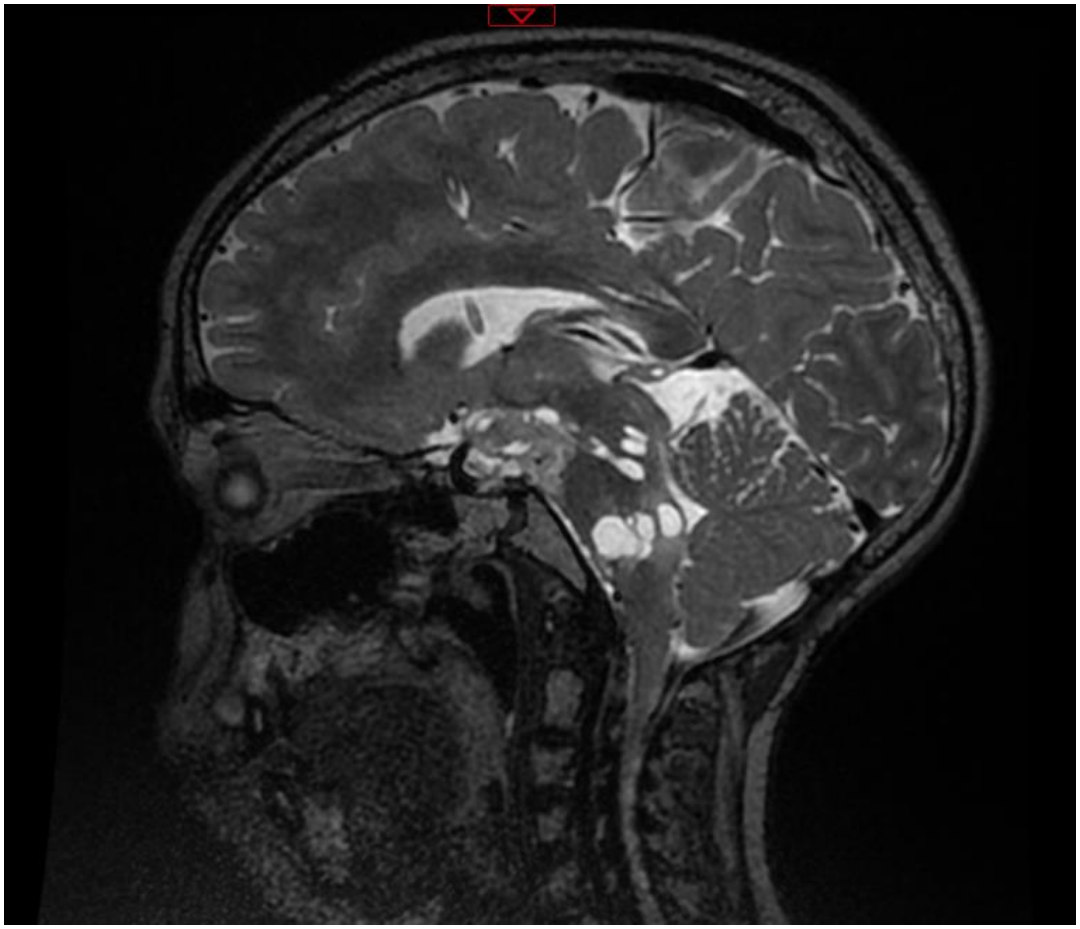
Total face mask (Philips)

Case 2

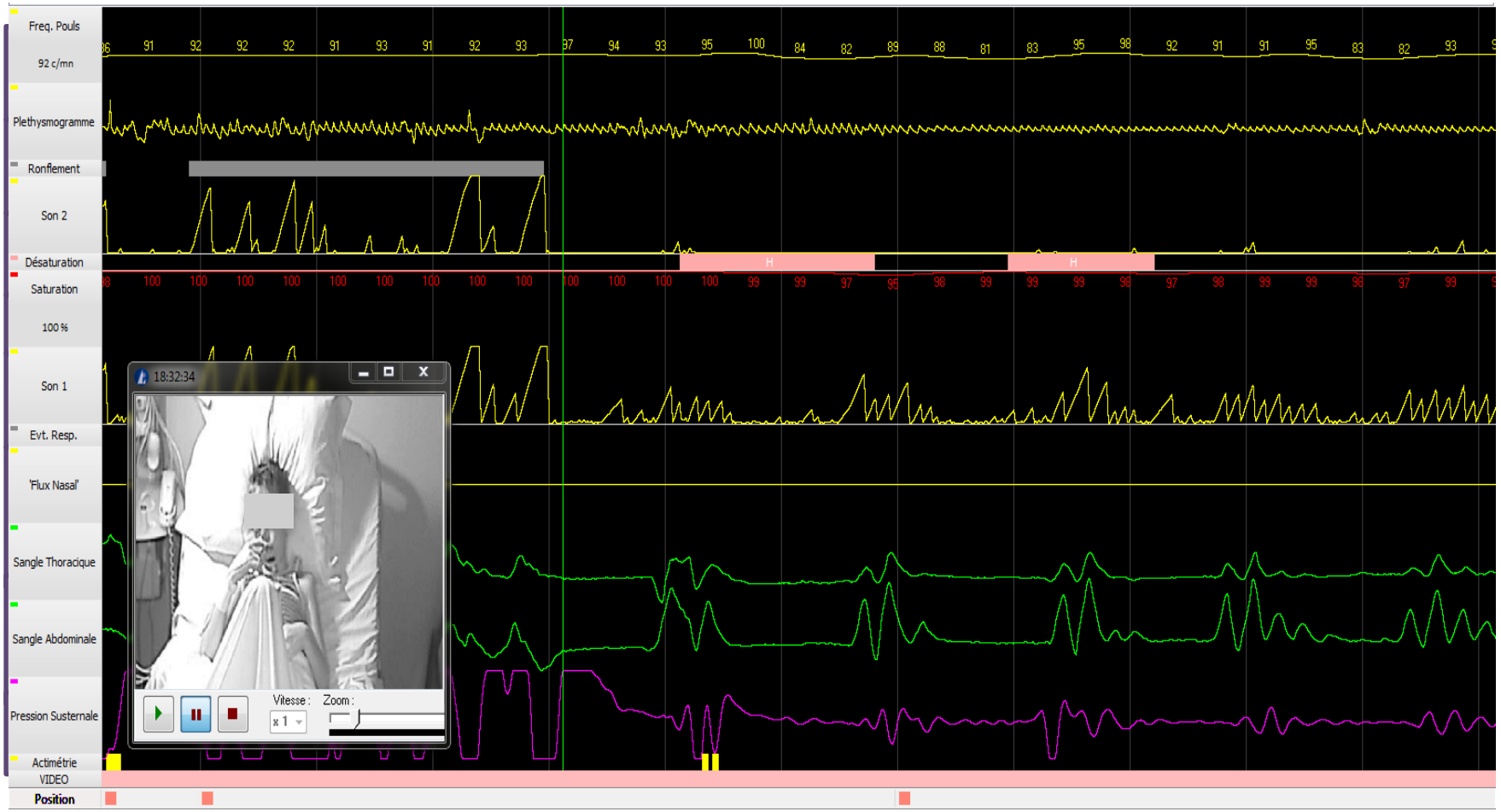
10-yr old boy with ganglioglioma

- Cerebral tumour diagnosed at age 8 yrs
- Numerous surgical interventions, hydrocephalus (ventriculo-peritoneal derivation), epilepsy, hypopituitarism substituted with L-thyroxine and hydrocortisone
- Sleep study because of daytime sleepiness

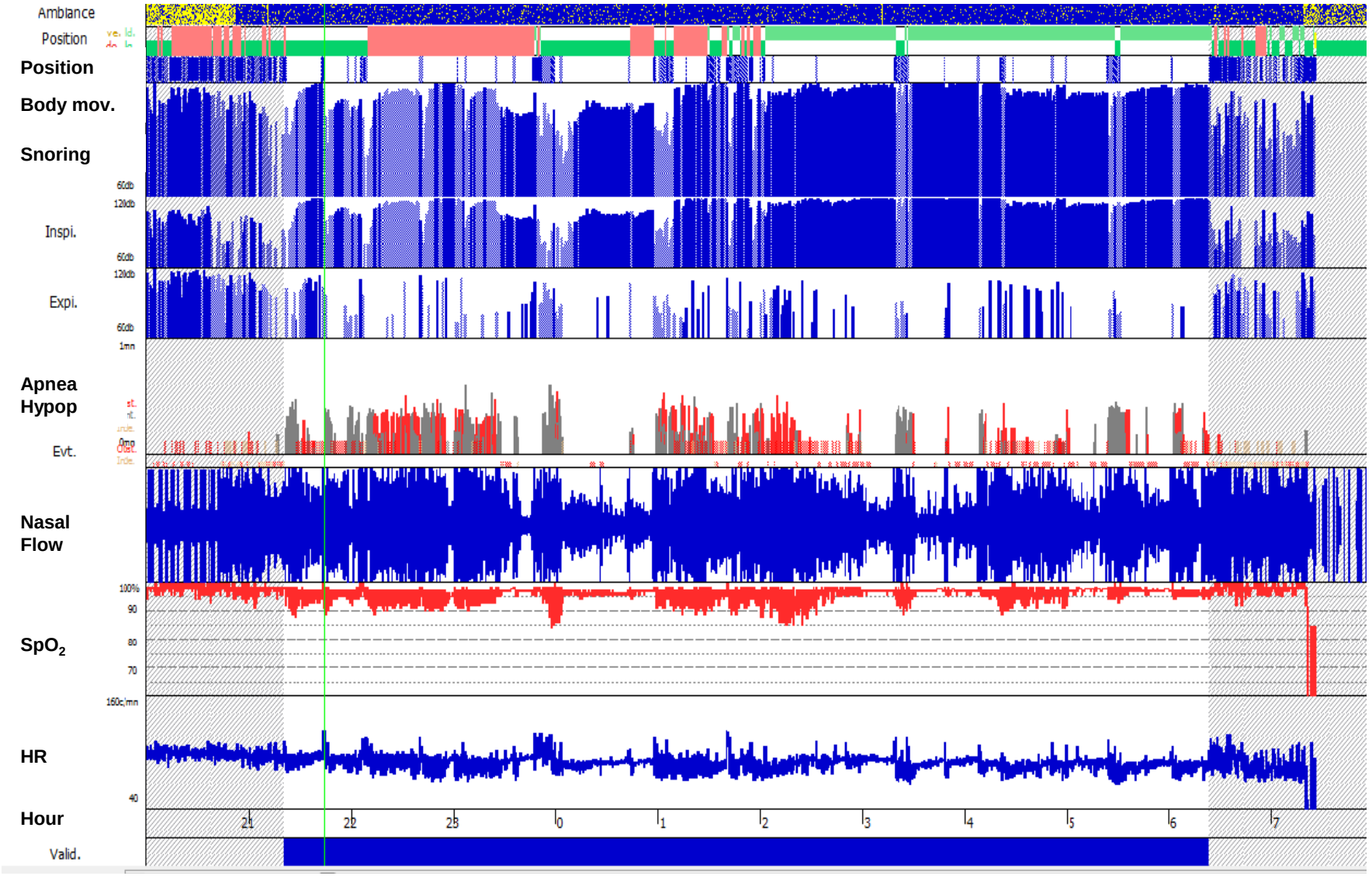
10-yr old boy with ganglioglioma



During wakefulness



Polygraphy


Ambiance
Position
Position
Body mov.
Snoring
60db
120db
Inspi.
60db
120db
Expi.
60db
1mn
Apnea
Hypop
Evt.
Nasal
Flow
100%
90
80
70
SpO2
160c/mn
40
HR
Hour
21
22
23
0
1
2
3
4
5
6
7
Valid.

Polygraphy


Durée Page
20 s
30 s
1 min
2 min
5 min
10 min
Voies
Tendances
Créer un Evénement
Evénements
Evt. Resp.
Page
Heart rate
PWA
Snoring
SpO2
Tracheal sound
Nasal flow
Thoracic belt
Abdominal belt
Sternal pressure
Position

Polygraphy


Durée Page
20 s
30 s
1 min
2 min
5 min
10 min
Voies
Tendances
Créer un Evénement
O
C
M
E
J
N
H
Evènements
Evt. Resp.
Page
Heart rate
PWA
Snoring
SpO2
Tracheal sound
Nasal flow
Thoracic belt
Abdominal belt
Sternal pressure
Position

Polygraphy


Heart rate
PWA
Snoring
SpO2
Tracheal sound
Nasal flow
Thoracic belt
Abdominal belt
Sternal pressure
Position

AHI 104/h (CAHI 50/h and OAHI 54/h) with mean $PtcCO_2$ 48 mmHg What is your attitude ?

1. Oxygen therapy
2. Bilevel ventilation
3. CPAP
4. Nothing, the disease is evolutive

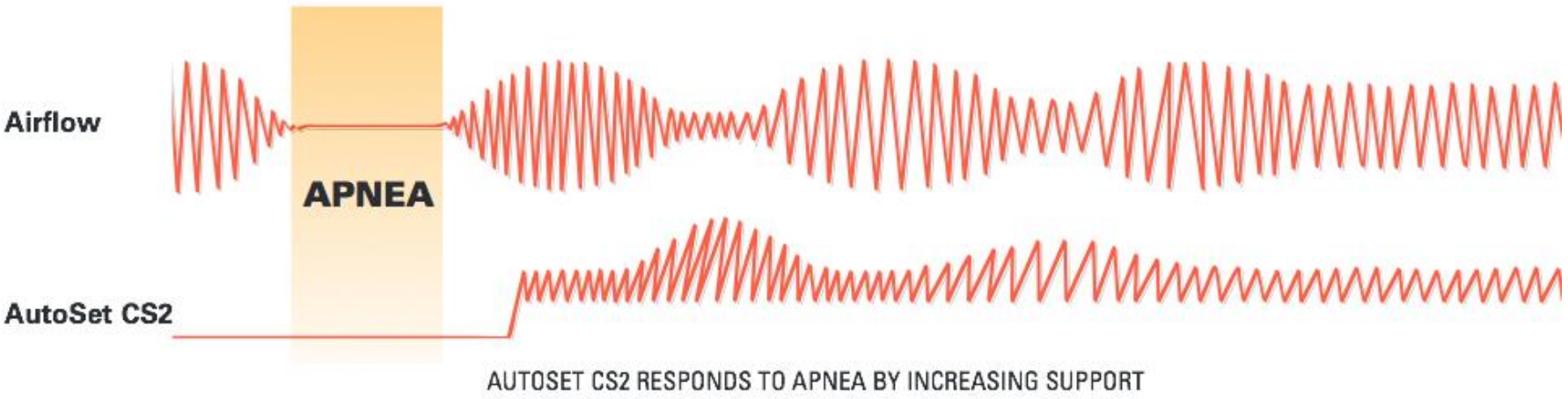
AHI 104/h (CAHI 50/h and OAHI 54/h) with mean $PtcCO_2$ 48 mmHg What is your attitude ?

1. Oxygen therapy
2. Bilevel ventilation
3. CPAP
4. Nothing, the disease is evolutive

Despite several attempts, the child did not tolerate CPAP neither Bilevel ventilation What is your attitude ?

1. Oxygen therapy
2. Autotitrated CPAP
3. Nothing
4. Other ?

Assisted servoventilation (ASV)



ASV delivers servo-controlled inspiratory pressure support on top of positive expiratory airway pressure

ASV beyond Cheyne Stokes

BMJ Case Reports

Novel treatment (new drug/intervention; established drug/procedure in new situation)

Chiari malformation and central sleep apnoea: successful therapy with adaptive pressure support servo-ventilation following surgical treatment

Ahmed Fahim, Anthony OC Johnson

57 yrs old woman with Chiari malformation

Table 1 Sleep studies during the course of investigation and management of the patient

Measure	Baseline	Auto SV	Auto SV postoperation	No NIV postoperation
AI	68	0.1	0.1	8.4
AHI	81	10.5	0.7	22.1
Total apnoeas	523	1	1	81
Central	522 (99%)	1 (100%)	1 (100%)	69 (85%)
Obstructive	1 (0%)	0	0	3 (4%)
Mixed	0	0	0	9 (11%)
Hypopnoeas	103	105	7	132
Duration mean (max)	23 (46)	16 (48)	14 (18)	17 (59)
Mean SpO_2	91%	95%	94%	93%
4% Dip rate	79	35	9.6	39.5

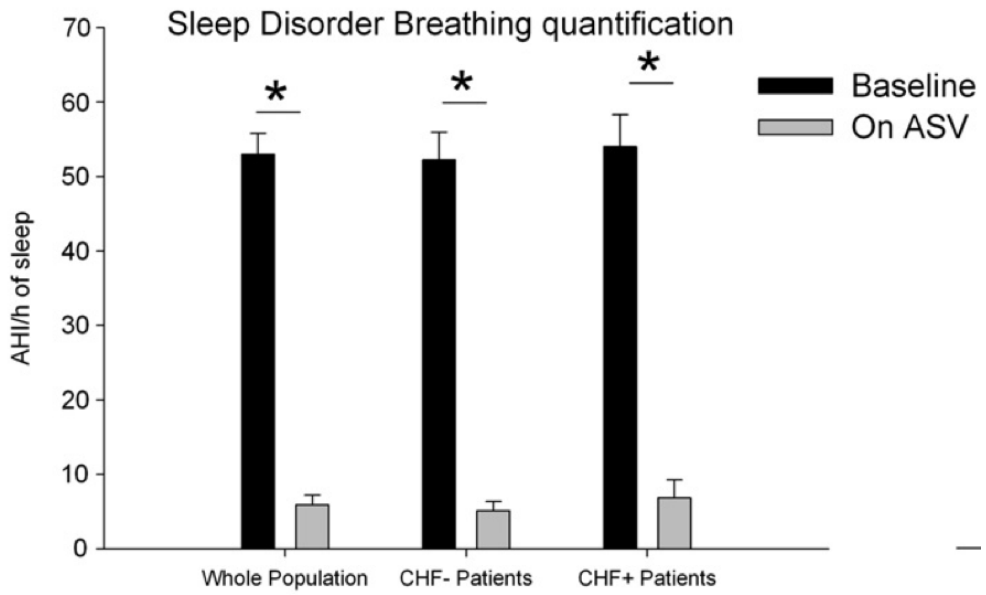
AHI, apnoea hypopnea index; AI, apnoea index; NIV, non-invasive ventilation; SV, servo-ventilation.

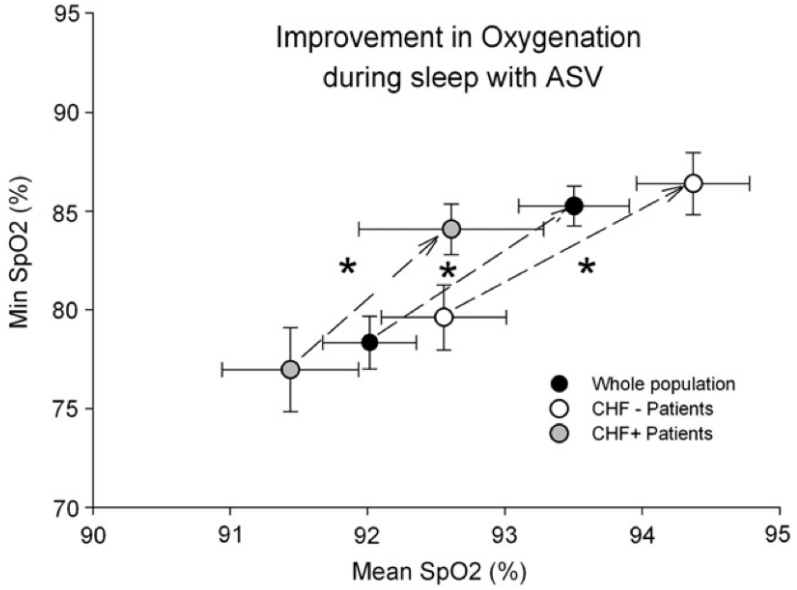
ASV beyond Cheyne Stokes

Effectiveness of Adaptive Servo Ventilation in the treatment of hypocapnic central sleep apnea of various etiologies

Claudio Carnevale [a,b,d], Marjolaine Georges [c], Claudio Rabec [c], Renaud Tamisier [a,b], Patrick Levy [a,b,1], Jean-Louis Pépin [a,b,1,*]

Sleep Medicine 12 (2011) 952–958



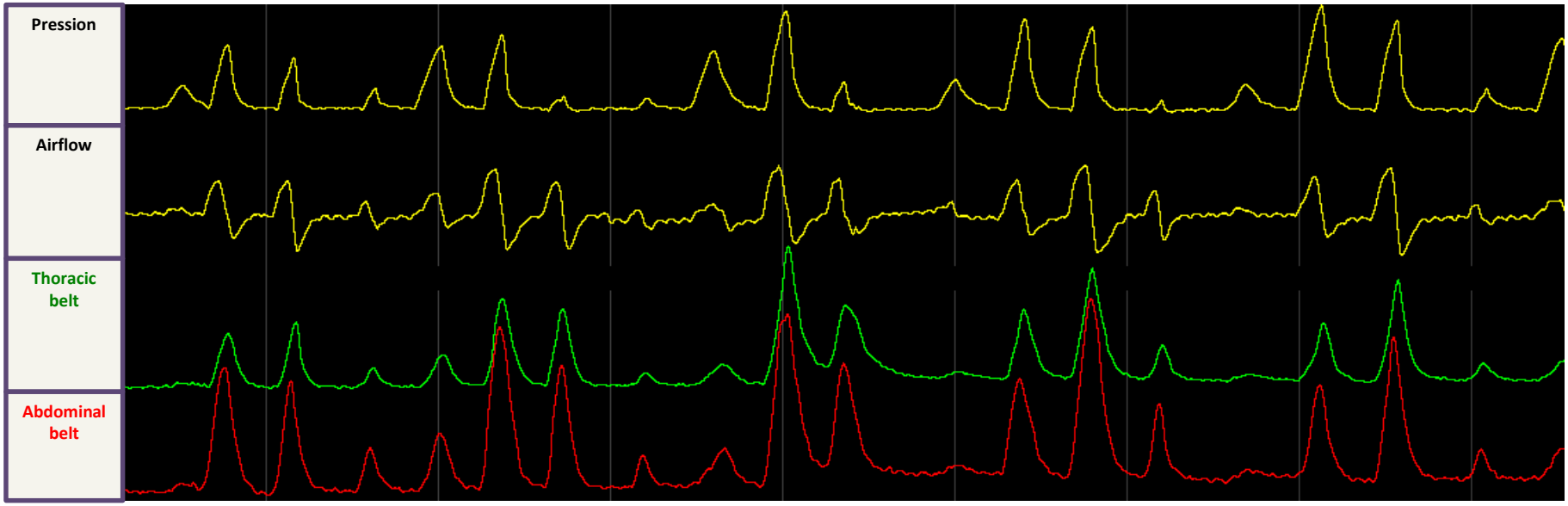


Our choice

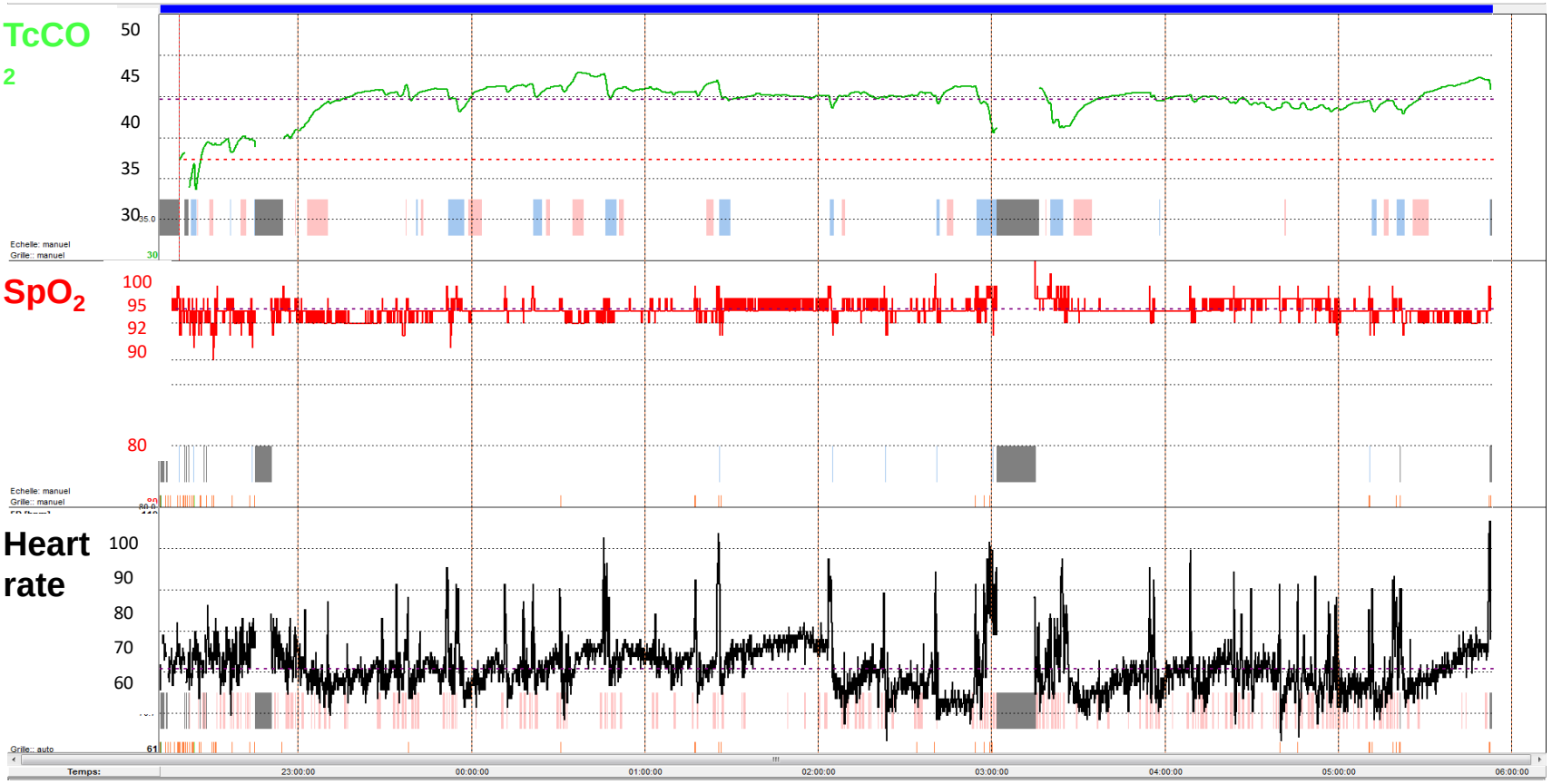
1. Ventilator: Resmed AirCurve 10 ASV
2. Interface: nasal mask
3. Pressure: EPAP 5-8 and IPAP 0-8 cmH_2O
4. Monitoring: polygraphy

Polygraphy during ASV

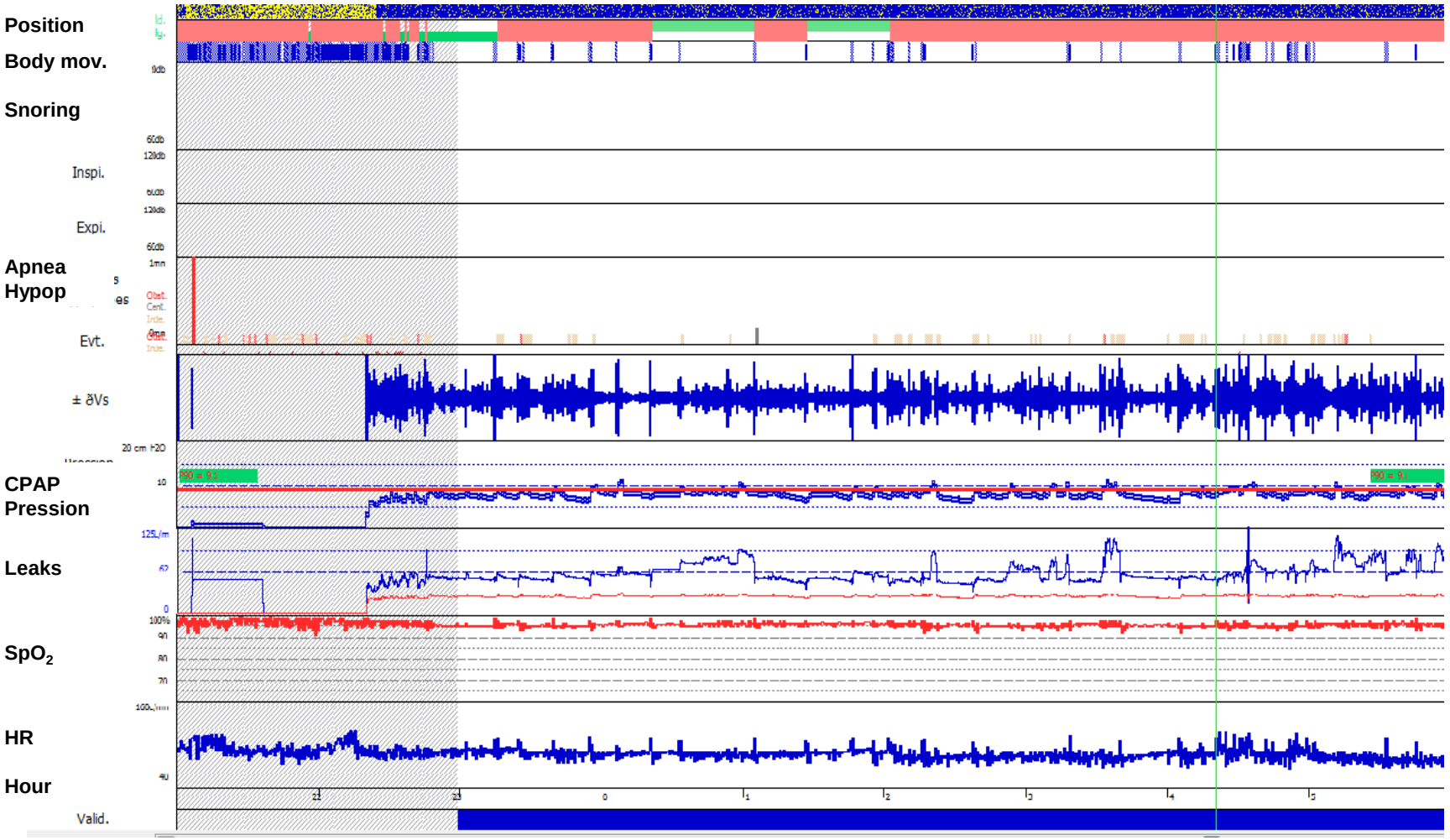
Polygraphy during ASV



Gas exchange during ASV



Polygraphy during ASV



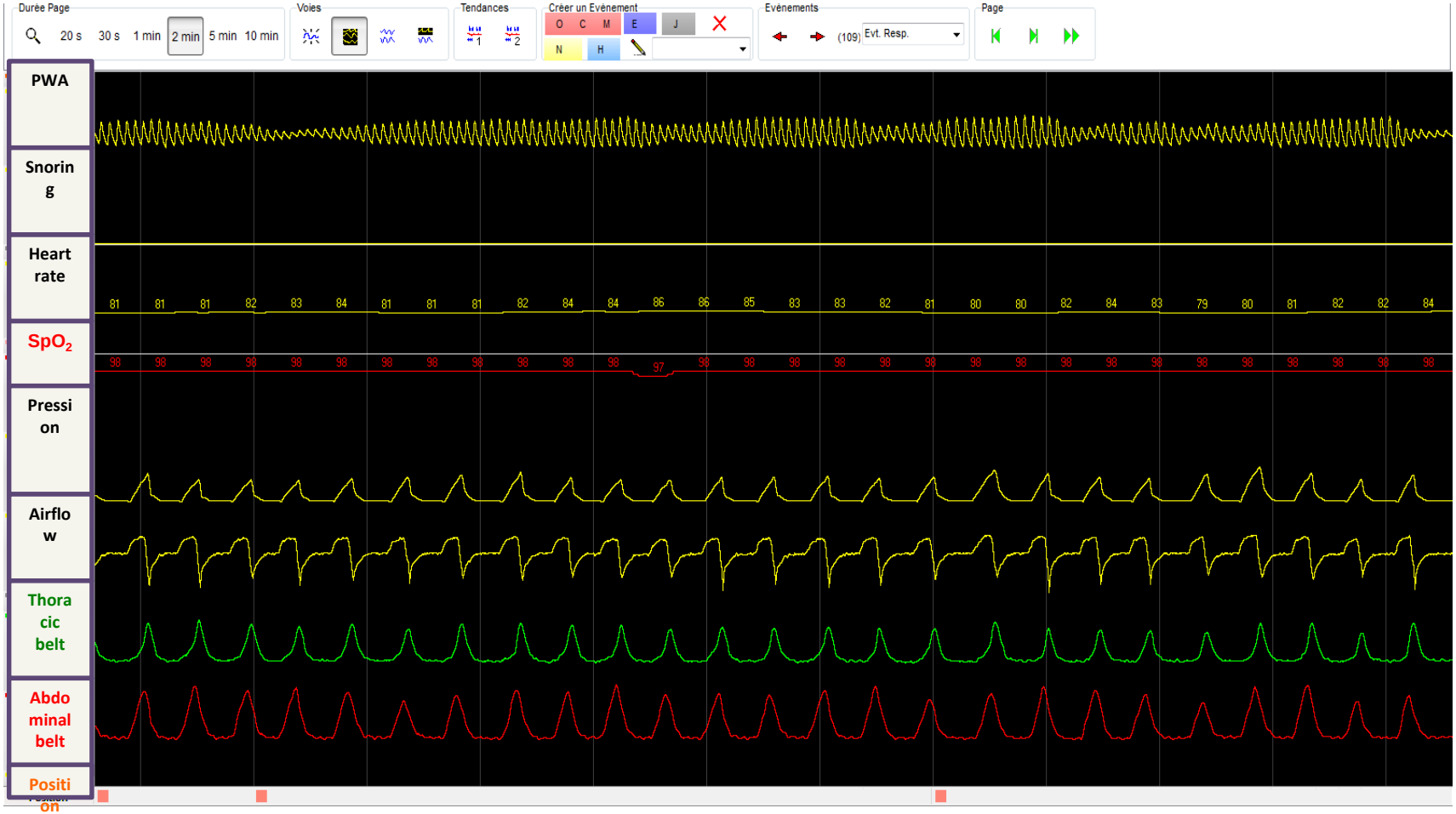
Gas exchange improved but sleep study is still abnormal, what would you do?

1. Switch to Bilevel ventilation
2. Increase EPAP
3. Increase IPAP
4. Nothing

Our choice

Increase IPAP from 0-8 to 3-8 cmH_2O

Polygraphy during ASV



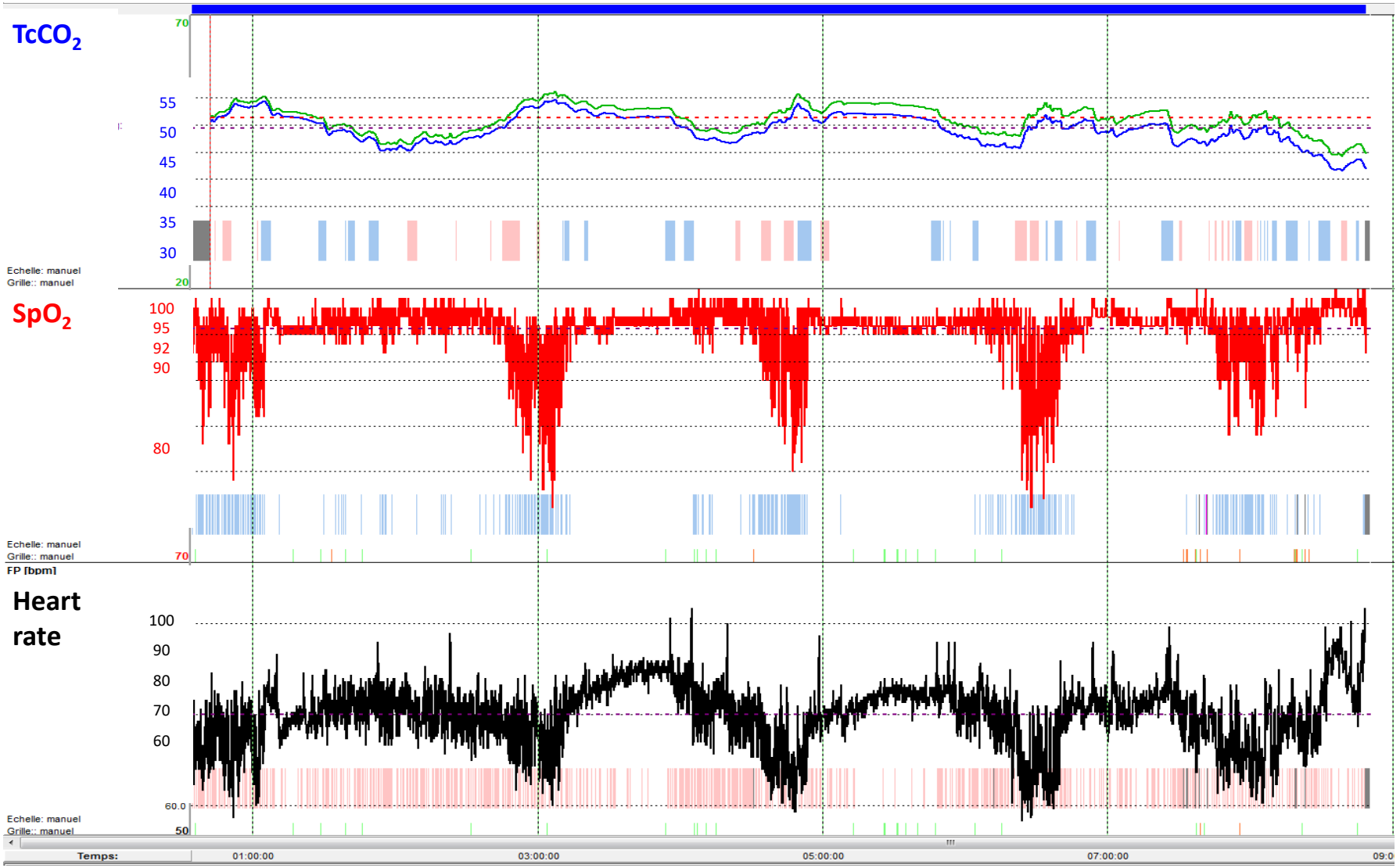
Polygraphy during ASV

Case 3

15-yr old girl with lymphoproliferative disease

- Lymphoid tissues hyperplasia with upper airway obstruction
- Obesity: BMI 37 kg/m^2
- Restless sleep with snoring

Nocturnal gas exchange



What would you do ?

1. Start CPAP
2. Start NIV
3. Perform a full PSG
4. Consult ENT surgeon

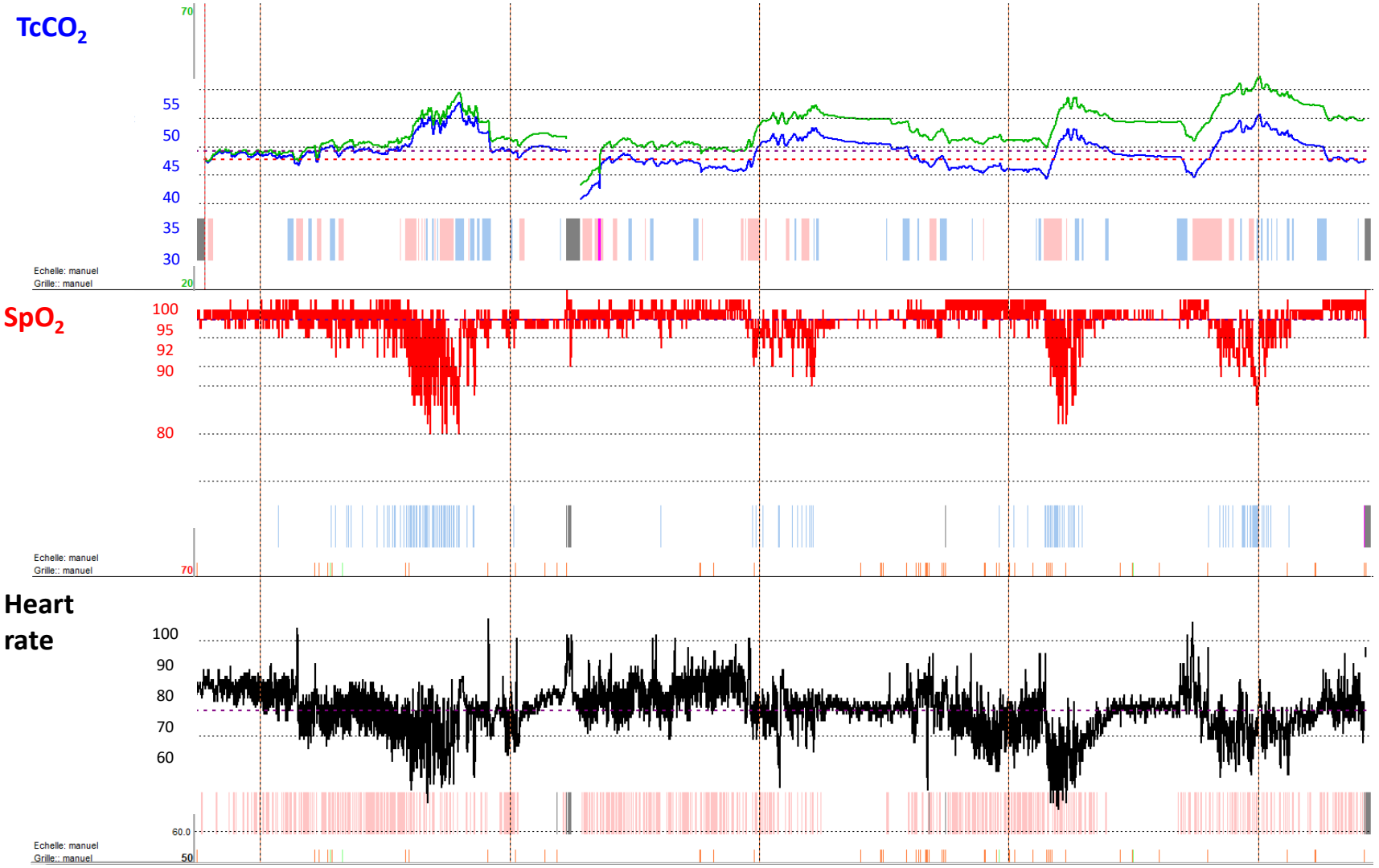
ENT surgery could not be done due to her underlying disease. What would you do ?

1. Start CPAP
2. Start NIV
3. Perform a full PSG

Our choice

1. Ventilator: Respironics BiPAP A40
2. Interface: facial mask (mouth breathing)
3. Settings: CPAP 10 cmH_2O
4. Monitoring of nocturnal gas exchange

Gas exchange during CPAP 10



Gas exchange showed persistant desaturation with concomitant hypercapnia at CPAP 10 cmH_2O. What would you do next ?

1. Increase CPAP pressure
2. Try autoset-CPAP
3. Switch to NIV

Clinical Guidelines for the Manual Titration of Positive Airway Pressure in Patients with Obstructive Sleep Apnea

Positive Airway Pressure Titration Task Force of the American Academy of Sleep Medicine

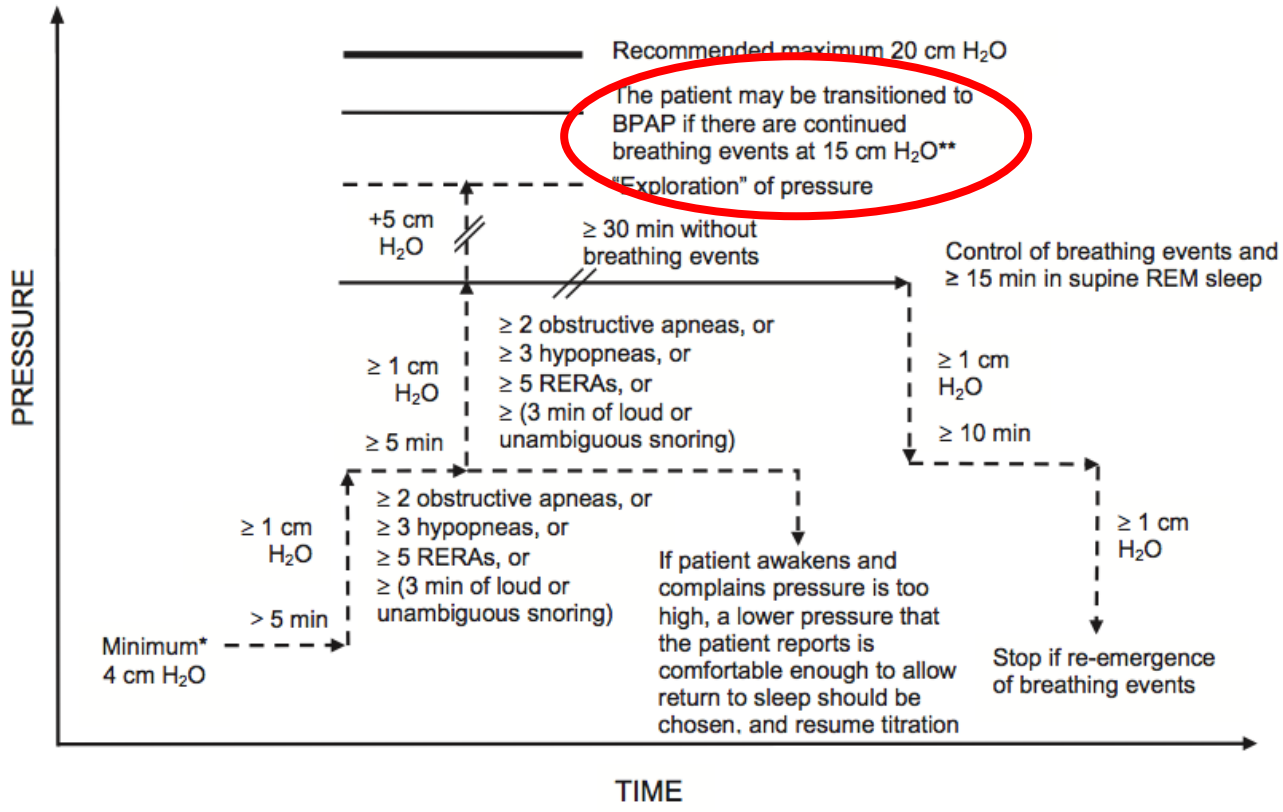
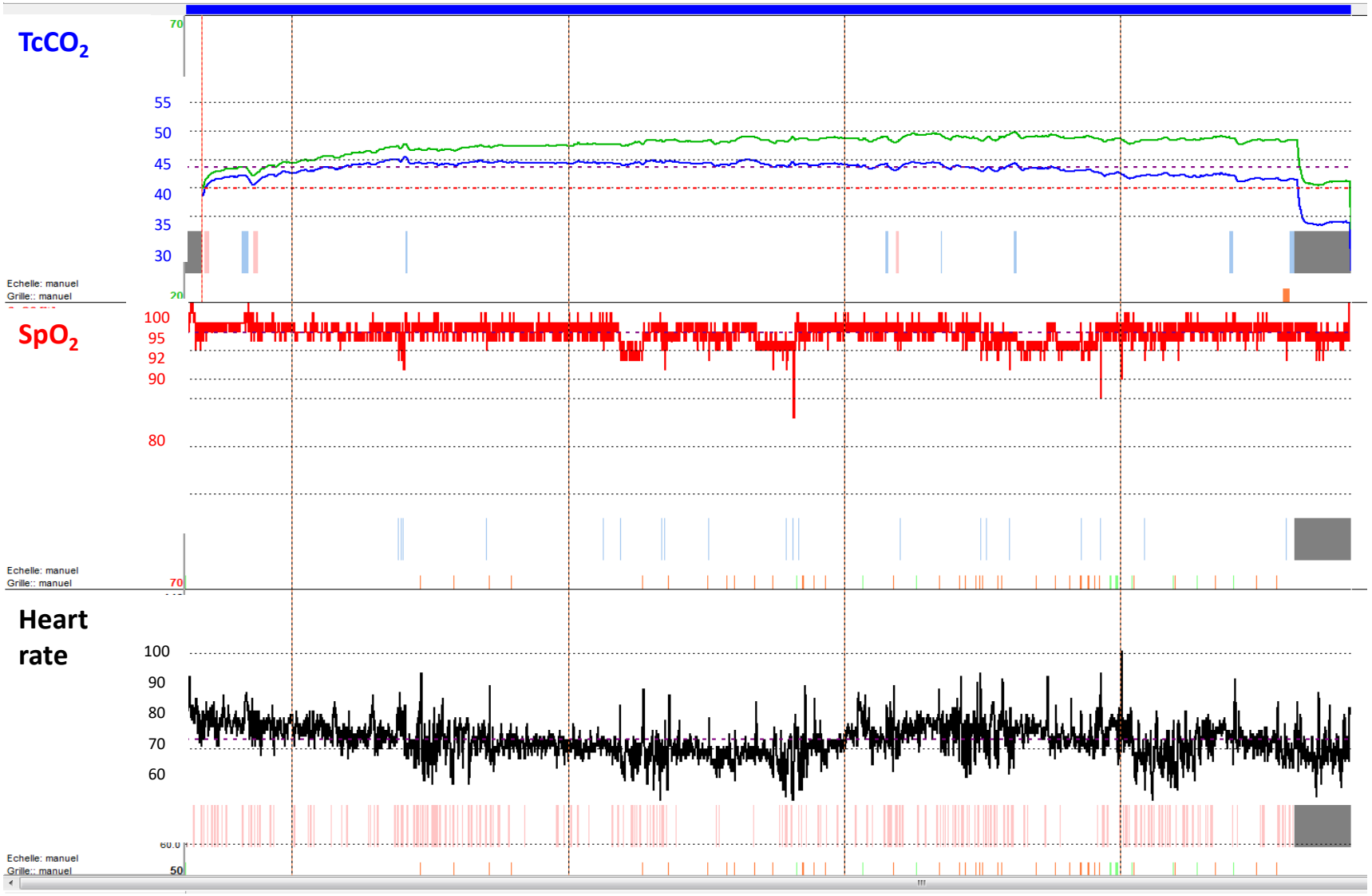


Figure 2—CPAP Titration Algorithm for Patients ≥12 years Dur-

But this girl was also obese and did hypoventilate so…we started NIV:

1. Ventilator: Respironics BiPAP A40
2. Interface: facial mask (mouth breather)
3. Settings: S/T AVAPS, IPAP 20/16 cmH_2O, EPAP 8 cmH_2O, VT 500 ml, RR 14 bpm, TgI 3 L/min TgE 20%

Gas exchange during NIV

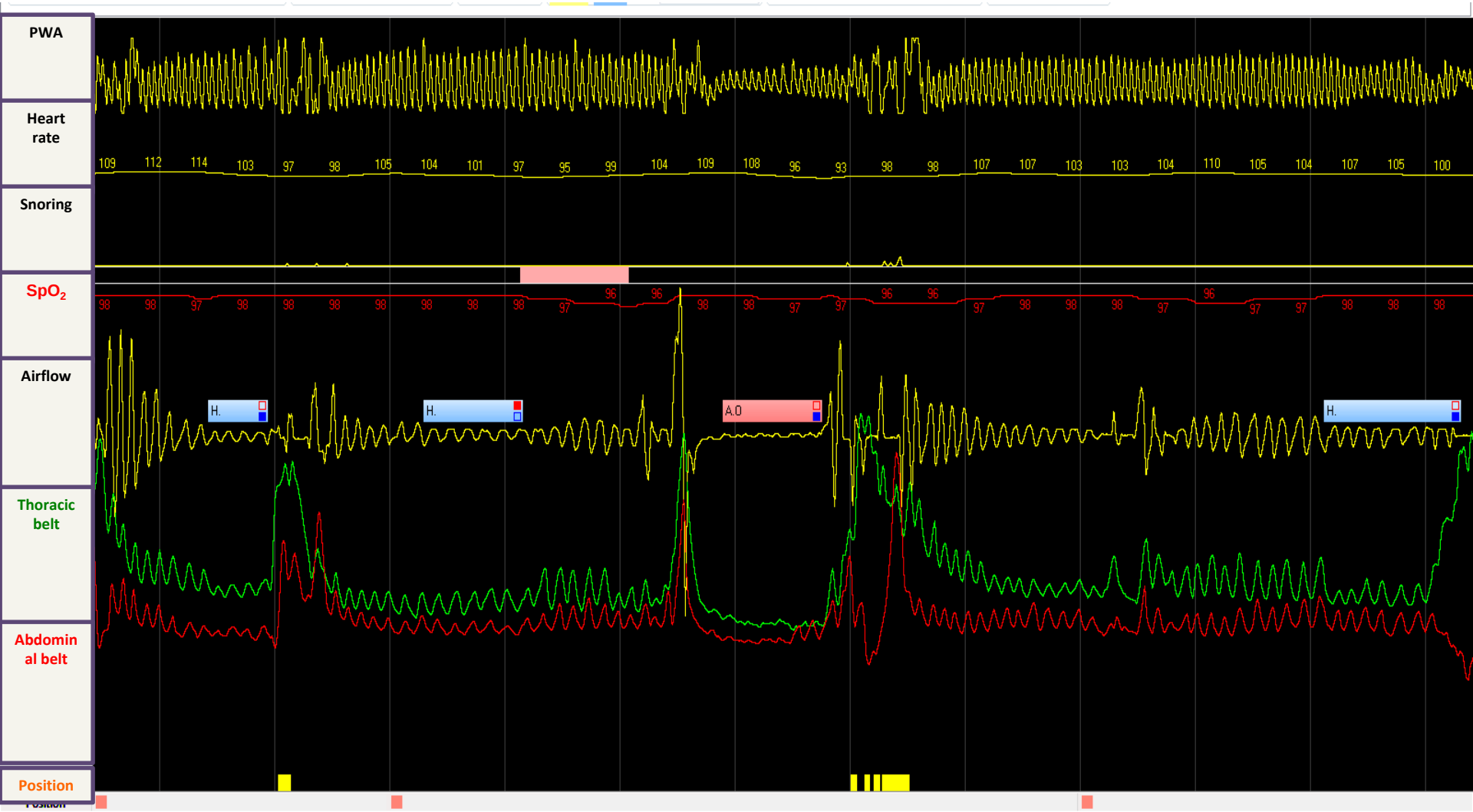


Case 4

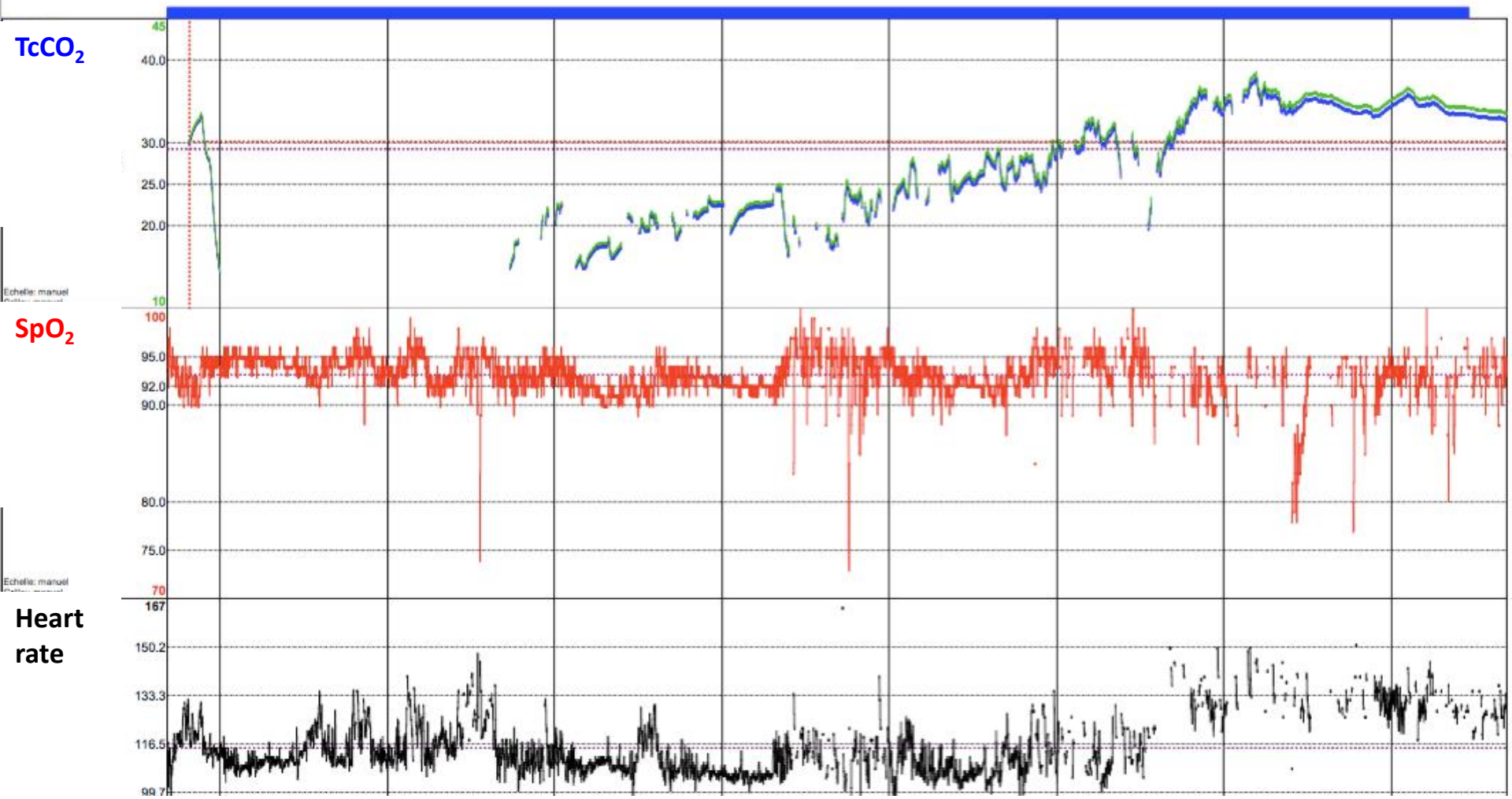
Paul, 18 months old, Down syndrome

- Congenital cardiopathy associated with pulmonary hypertension
- Restless sleep with snoring

Polygraphy: AHI 11/h



Nocturnal gas exchange



Severe OSA in a Down patient, what would you do?

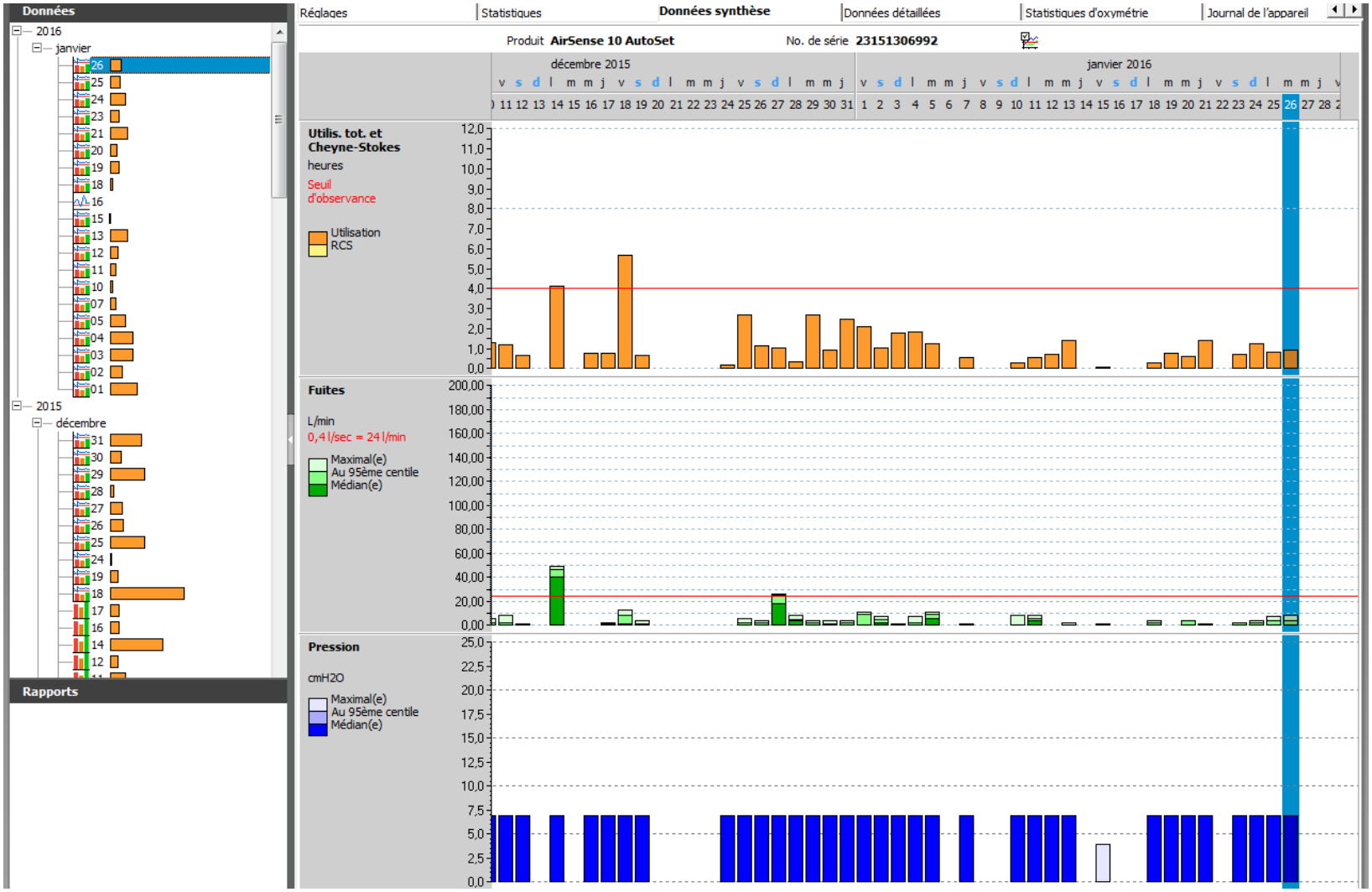
1. Start CPAP
2. Send to the ENT surgeon
3. Oxygen therapy
4. Nothing

- No indication of ENT surgery

Our choice

1. Ventilator: Resmed Stellar 150
2. Interface: Nasal mask
3. Setting: CPAP 7 cmH_2O
4. Monitoring of nocturnal gas exchange

CPAP was tolerated during the first 3 months, but then…



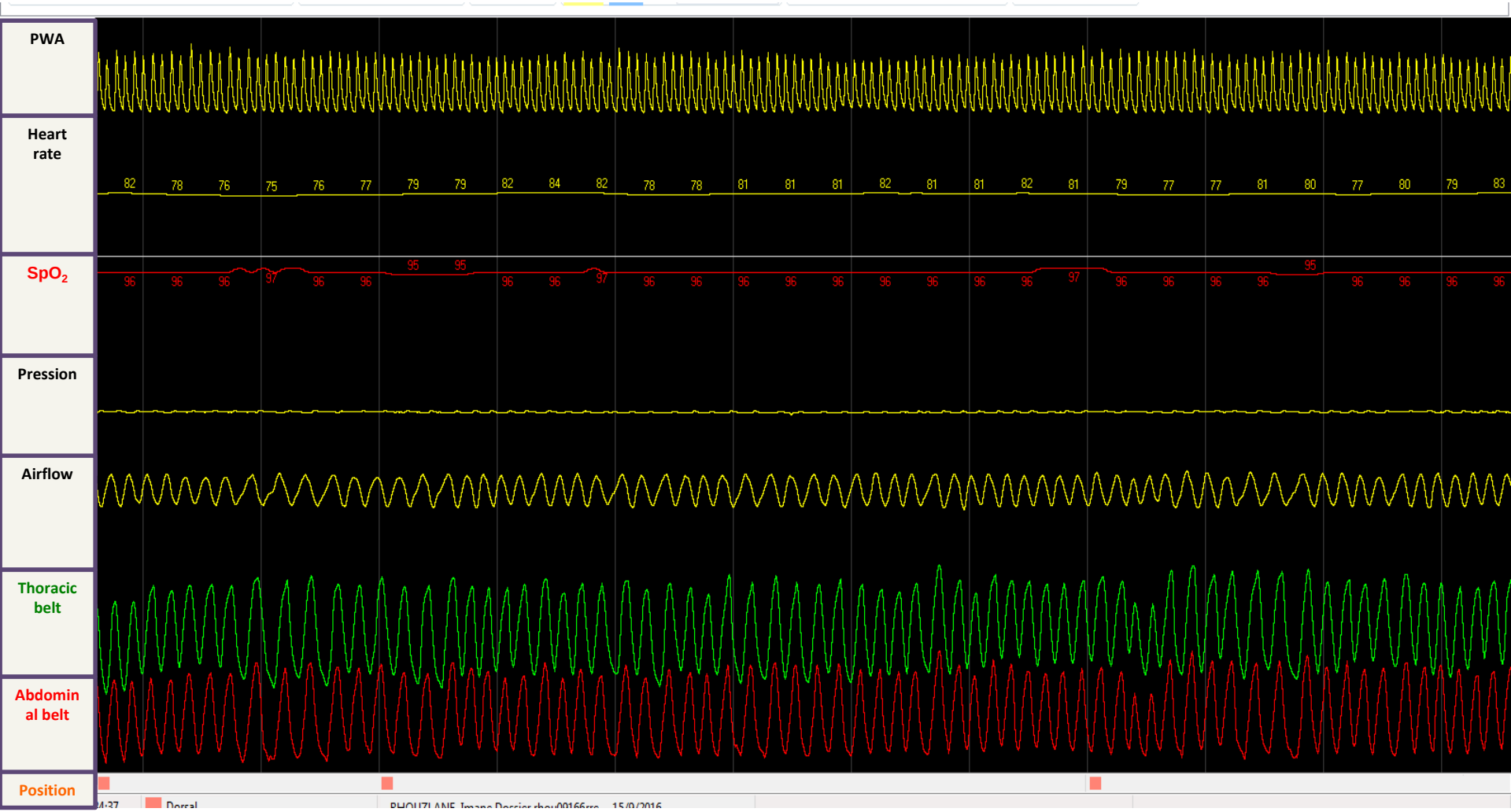
Poor CPAP tolerance, what would you do?

1. Do an overnight polygraphy
2. (Re)-send to the ENT surgeon
3. Nothing
4. Decrease the CPAP pressure
5. Switch to autotitrated CPAP
6. Switch to some other device

Our choice: high flow

1. Start with a flow of 2L/min in air
2. Cannula not completely obstructing the nares
3. Temperature 31˚ C
4. Sleep study to titrate the flow

Polygraphy under HFNC

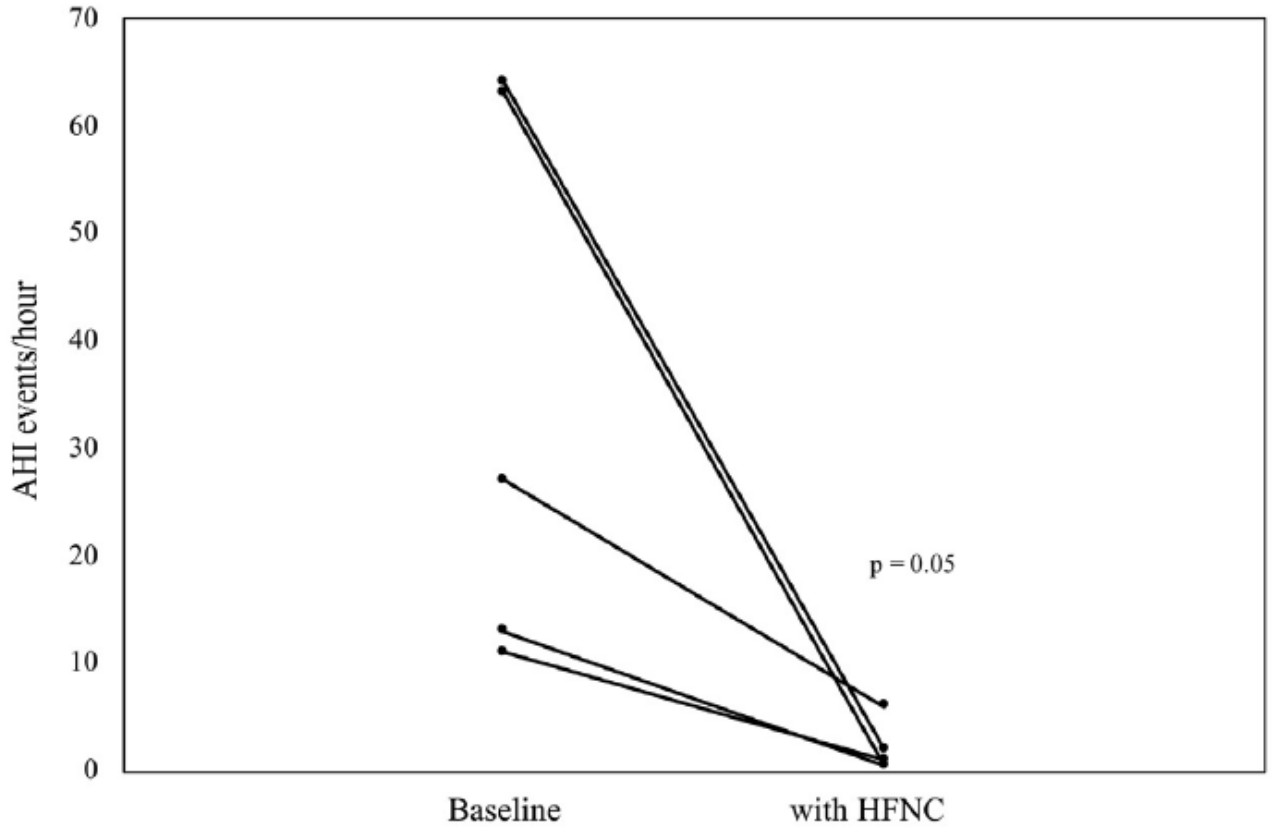


Paul tolerates HFNC and is doing well

- Mean HFNC use 7.3h/night
- Pulmonary hypertension stable

High-flow nasal cannula for children not compliant with continuous positive airway pressure

Alessandro Amaddeo [a, b, *], Sonia Khirani [a, b, c], Annick Frapin [a], Theo Teng [a], Lucie Griffon [a, b], Brigitte Fauroux [a, b]



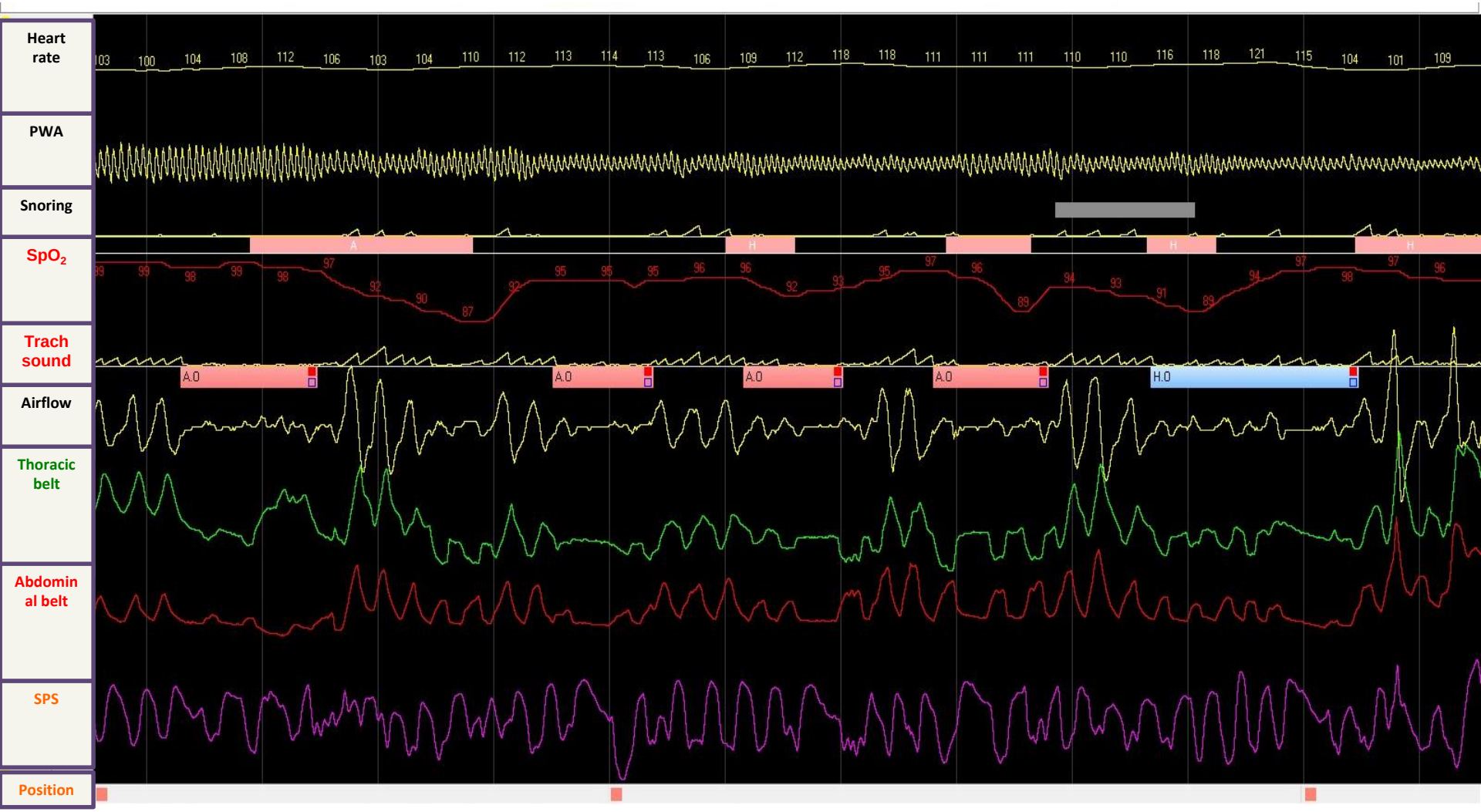
Sleep Medicine 63 (2019) 24–28

Case 5

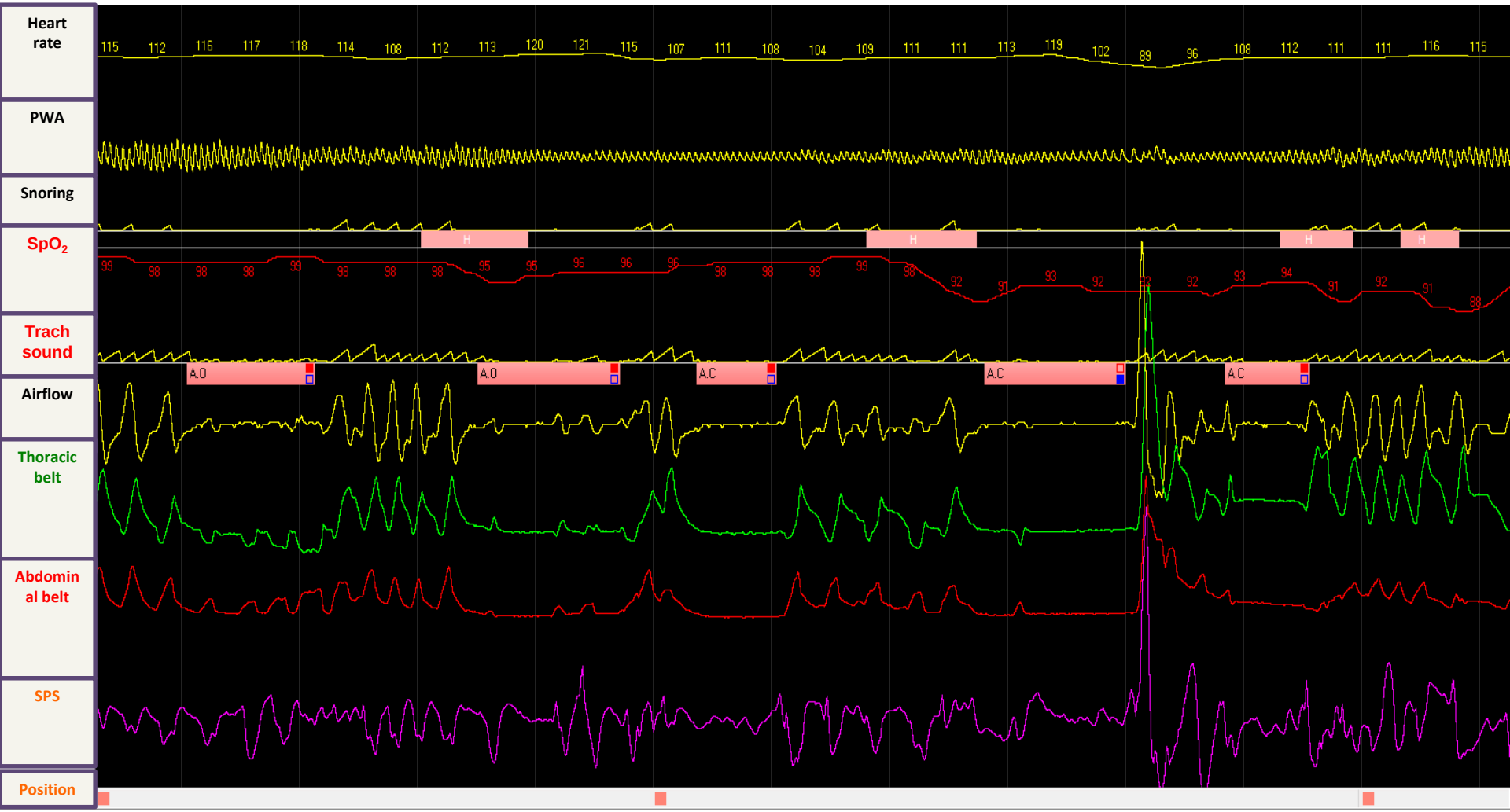
Baptiste, 22 months old, myelomeningocele and Chiari malformation

- Persistant nasal obstruction
- Snoring and apneas reported by the parents
- Restless sleep

Polygraphy: AHI 34/h



Polygraphy: AHI 34/h (CAI 2/h)



Severe OSA in a patient with myelomeningocele and Chiari malformation, what would you do?

1. CPAP
2. Send to the ENT surgeon
3. Oxygen
4. Send to the neurosurgeon

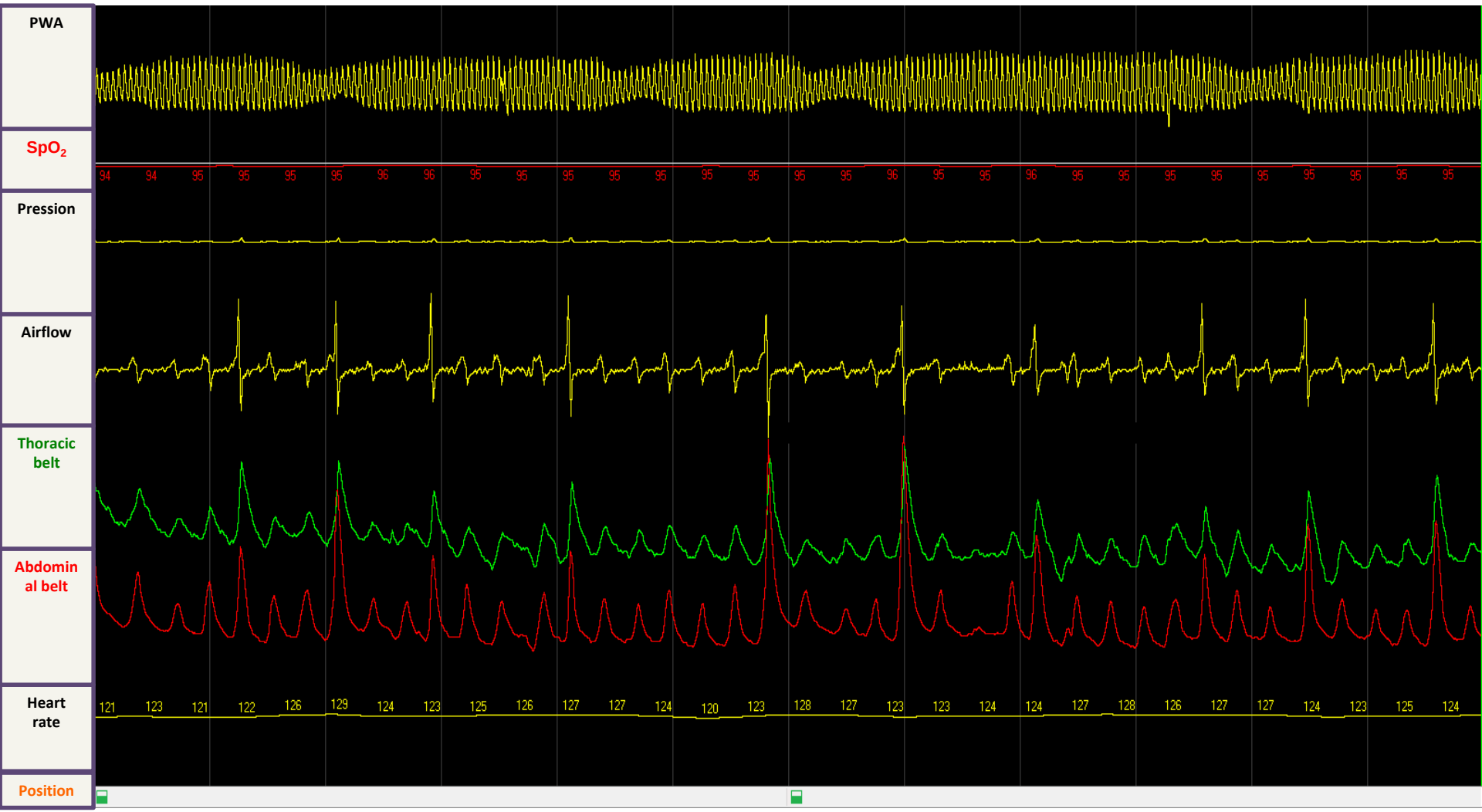
Severe OSA in a patient with mylomeningocele and Chiari malformation, what would you do?

1. CPAP
2. Send to the ENT surgeon: no indication for any upper airway surgery
3. Oxygen
4. Send to the neurosurgeon

Our choice

1. Ventilator: Resmed AirSense 10
2. Interface: nasal mask
3. Pressure: CPAP 8 cmH_2O
4. Monitoring : titration study

Titration study at 8 cmH_2O



Appareance of central apneas

Treatment-emergent central sleep apnea (TECSA) : appearance of central apneas and hypopneas after significant resolution of OSA with CPAP therapy

Risk factors for TECSA:

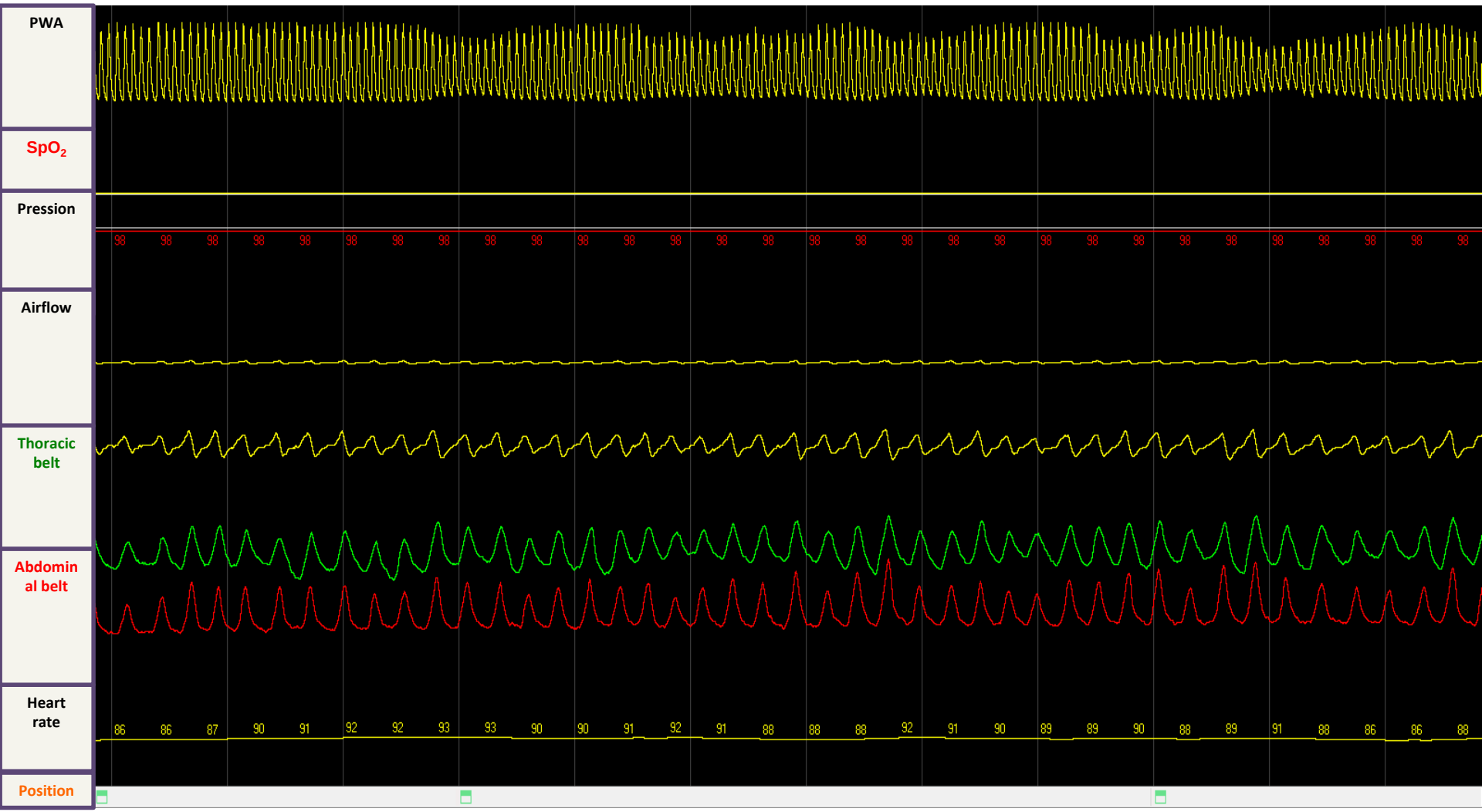
- Male gender
- Higher baseline AHI
- Higher baseline CAI

TECSA may occur at any CPAP level but is more likely to appear at higher level (CPAP > 10 cmH_2O)

TECSA in a patient with mylomeningocele and Chiari malformation, what would you do?

1. Start Bilevel ventilation
2. Decrease CPAP level
3. Maintain the same CPAP level
4. Send to the neurosurgeon

Polygraphy at 8 cmH_2O



The Significance and Outcome of Continuous Positive Airway Pressure-Related Central Sleep Apnea During Split-Night Sleep Studies*

CHEST / 132 / 1 / JULY, 2007

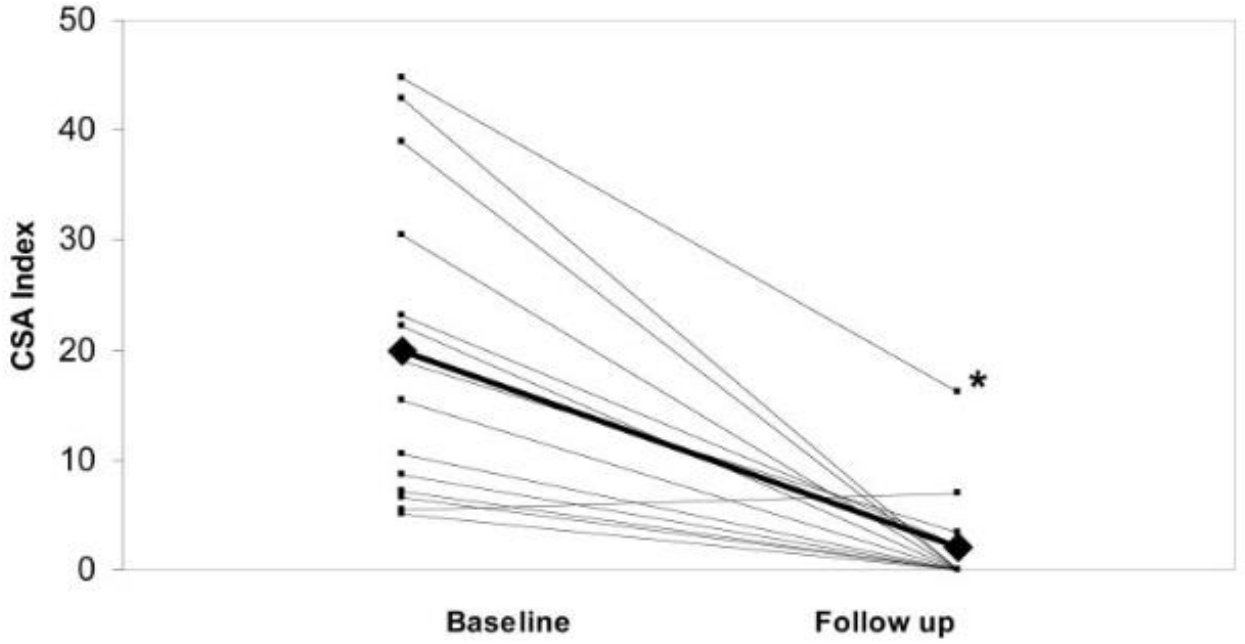


FIGURE 1. Change in CSAI in individual 14 subjects with CPAP-related CSA (diamond sign indicates mean values). *Subject with diastolic heart failure demonstrating persistent CSA on follow-up study.

Nearly all patients with CPAP-induced CSA at baseline had complete resolution of the central events on follow-up when studied on the prescribed level of CPAP used to eliminate OSA.