Interpretation of Pediatric Pharyngeal Manometry

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Objectives: Interpretation of Pediatric Pharyngeal Manometry

- Identify abnormalities that may be identified during pediatric HRPM which may not be detected on other instrumental assessments
- Review case studies, interpretations, and subsequent recommendations
- Describe potential for monitoring therapeutic efficacy

Pediatric Manometry Literature

- Limited to a handful of research groups over the past few decades as the technology has advanced
- No normative reference ranges exist for pediatrics
- Studies have generated data specific to conditions – velocardiofacial syndrome, rumination, muscular dystrophy, cerebral palsy
- Pharyngeal manometry in pediatric dysphagia assessment – review article summarizing current evidence
  - Ferris & Omari, 2019

HRM with Combined Impedance in Pediatrics

- Rommel, et al., 2015
- n = 35, median age 10.5 y
- Reported as first time HRM with combined impedance performed in children
- Children with post-fundoplication dysphagia found to have different motor responses to bolus movement compared to the other participants; shorter time between esophageal maximal distension and peak bolus pressure – indicating more pressurized bolus traveling through esophagus
- Discriminates the cause of dysphagia -*can differentiate paediatric patients with dysphagia symptoms in relation to either weak peristalsis (poor bolus clearance) or over-pressurization (abnormal bolus flow resistance).*

Use of HRM with impedance in pre and post fundoplication patients

- Omari et al., 2018
- n = 13, median age = 6.8 years
- Findings: patients reported symptoms of dysphagia preoperatively; fundoplication decreased dysphagia in most; however, some had postoperative dysphagia and were distinguishable by elevated bolus clearing pressures

Velocardiofacial Syndrome - VCF

- Rommel et al., 2008
- Case series VCF ages 1-3
- HRM + simultaneous VSS
- HRM showed "*subtle distinction between pathophysiology and characterization of pharyngeal and UES dysfunction*"
- Tailored interventions: UES dilation, UES botox, NPO rec

- AKA 22q11deletion syndrome, Shprintzen syndrome, DiGeorge
- Common features: cleft palate, heart defects, characteristic facial appearance (elongated face, almond shaped eyes, wide nose, small ears)

Rumination – use of HRM to confirm dx

- Grunder et al., 2017
- Rumination syndrome – expulsion of gastric content without retching, pain, nausea, vomiting
- n = 15, < 18 y
- HREM confirmed diagnosis and pathophysiology – provided as explanation
- *Further research needed to determine whether results influence treatment and outcomes for children with rumination syndrome

Pressure Flow Metrics and Abnormal Dysphagia Disorders Survey/Functional Oral Intake Scale (FOIS)

- Ferris et al., 2016
- n = 45, median age = 5 y; controls = 34
- Heterogeneous population – neurologic, metabolic, tracheostomy patients with dysphagia
- Between patients and controls – key differences in UES resistance and UES distension found, consistent with reduced UES relaxation and opening in patients with oropharyngeal dysfunction
- Objective profiling of bolus timing and efficiency of bolus clearance – suggest greater prevalence of UES dysfunction which could be targeted in treatment

Achalasia in Children

- Singendonk, et al., 2018
- n = 20 – aim was to evaluate esophageal motor function in children diagnosed with achalasia
- Used novel esophageal pressure flow variables – future studies recommended to correlate parameters with symptom severity before and after therapy

Defining Pharyngeal Contractile Integral during HRM in Neonates

- Jadcherla et al., 2018
- n= 19; neonates born between 34-41 weeks (MA=38.6) gestation evaluated at 42.9 weeks with HRM
- Characterization of pharyngeal contractile integral (PhCI) in neonates – all pharyngeal regions examined
- PhCI of oral stimuli swallows were distinct from pharyngeal stimuli and spontaneous swallows
- Conclusion – *PhCI is a novel, metric capable of distinguishing (1) proximal and distal pharyngeal activity, (2) effects of oral and pharyngeal stimulation, and (3) effects of prolonged stimulation. Changes in pharyngeal contractility with maturation, disease, and therapies can be examined with PhCI.*

Inter- and Intrarater Reliability of the Chicago Classification in Pediatric High Resolution Esophageal Manometry Recordings – Singendonk et al., 2014

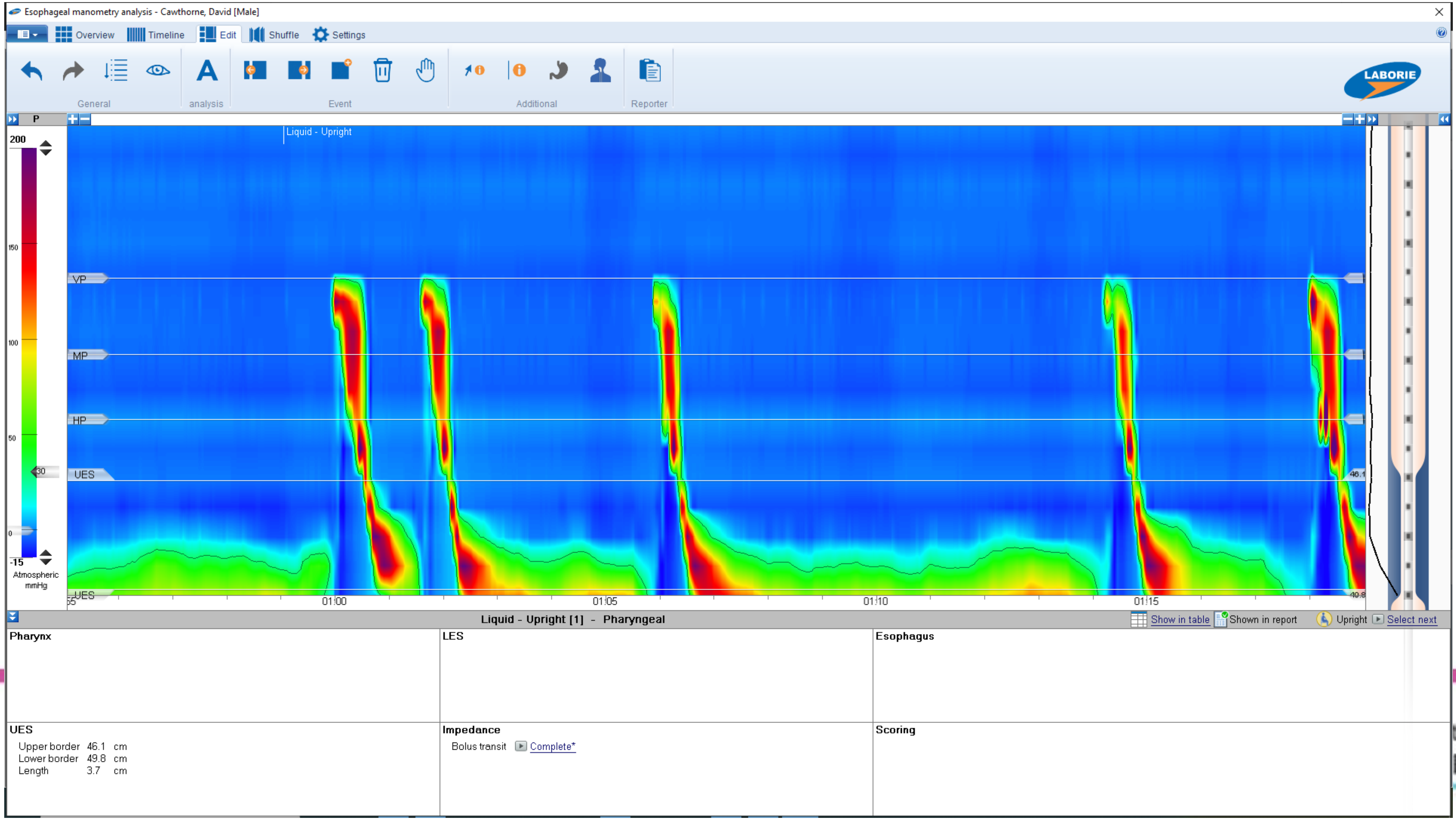
- n = 30
- Mean age: 12.1
- In pediatric studies, intra- and interrater reliability among experienced and inexperienced raters was found to be excellent for the esophageal pressure topography metrics ‘integrated relaxation pressure’, ‘distal contractile integral’, and ‘break size’
- Automated software-based Chicago Classification (CC) aids in the diagnosis of pediatric esophageal motility disorders with high inter- and intrarater reliability. However, the clinical diagnosis of achalasia and distal esophageal spasm is less reliable

Interpretation of Results

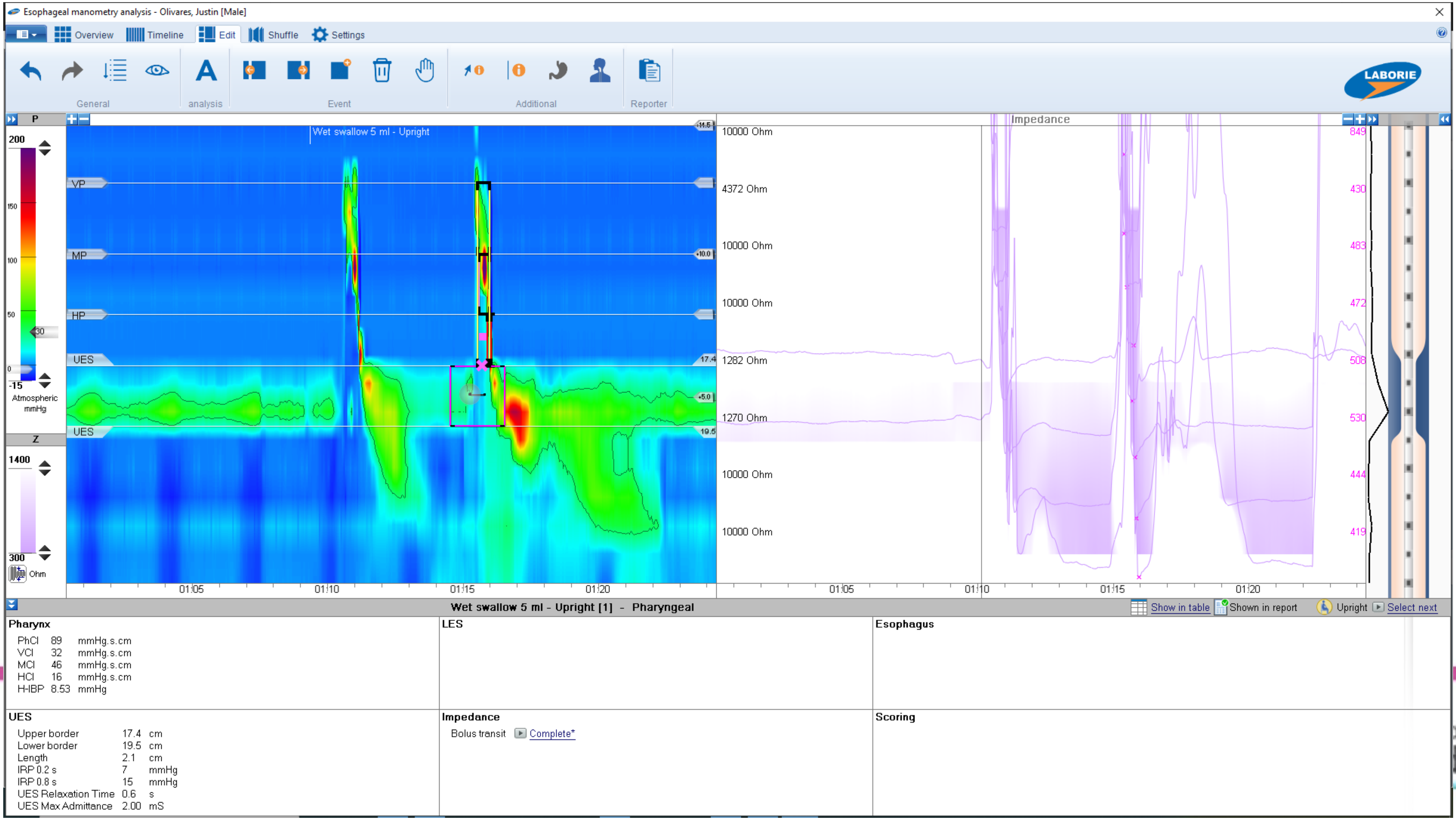
Interpretation of Results

- Allow software to identify swallows
- Often need to visually scan the tracings for appropriate swallows to analyze
  - Topography
  - Impedance
- We are currently comparing FEES results with HRPM results to allow calibration of our findings and interpretation of clinical significance of findings

Typical Pressure Topography



Impedance Overlay



Review of Our Data on the First 28 Patients

Table 6. FEES Parameters and Association with HRPM

Variable	PhCI	VCI	MCI	HCI	UES IBP	UES IRP 0.2 s	UES IRP 0.8 s	UES Max Admit (mS)	UES RT (s)
Oral motor abnormality						↑*			
Hypopharynx/Larynx abnormal									
Abnormal VF mobility	↓*	↓*	↓						
Abnormal secretion management		↑		↑		↑*	↑*		
Decreased or absent spontaneous swallow						↑*	↑		
Increased secretions in hypopharynx						↑*	↑*		
Impaired aspiration response	↓	↓*							
Decreased sensation					↑*	↑*	↑*		
Delayed or no swallow initiation		↓			↑	↑*	↑*		
Penetration identified						↑*	↑*		
Aspiration identified					↑				
Pharyngeal Residue									
Total n=28									

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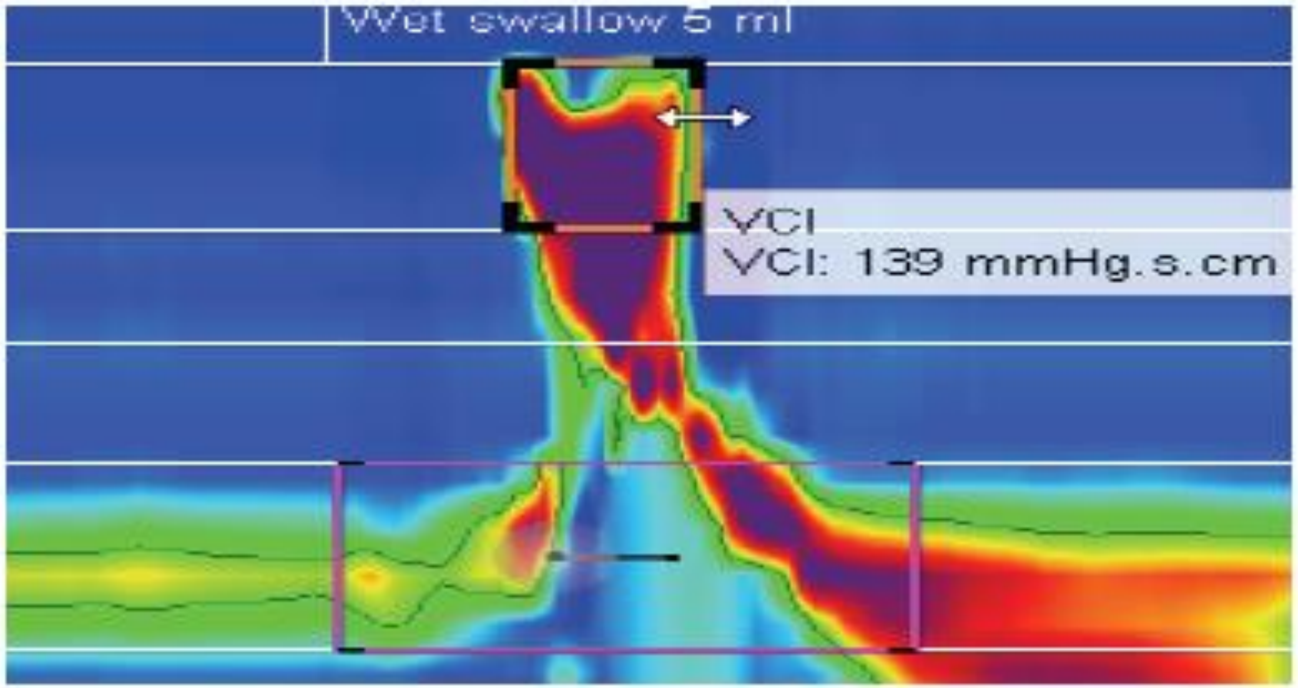
Velopharyngeal Contractile Integral (VCI)

Measure of contractile vigor within a space-time box on the pressure topography plot spanning the velopharyngeal region only

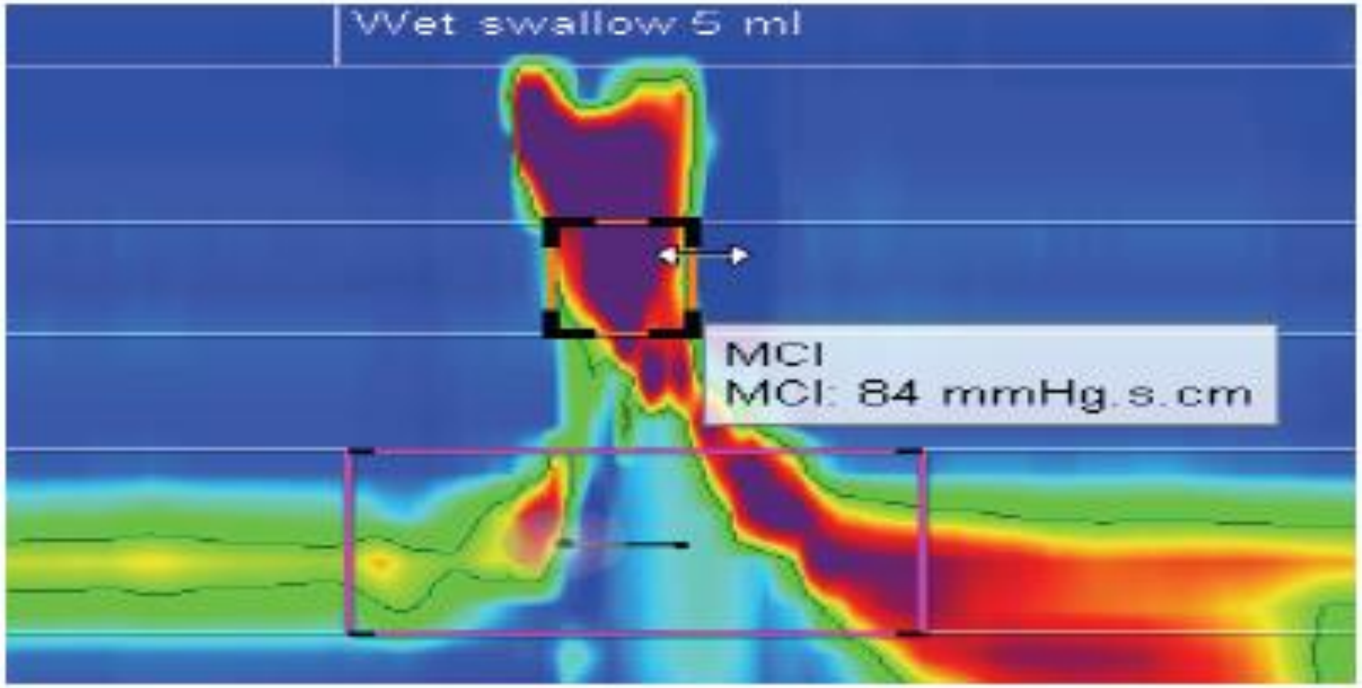
Mean pressure within this domain multiplied by duration (s) and length



Mesopharyngeal Contractile Integral (MCI)

Measure of contractile vigor within a space-time box on the pressure topography plot spanning the mesopharyngeal region only

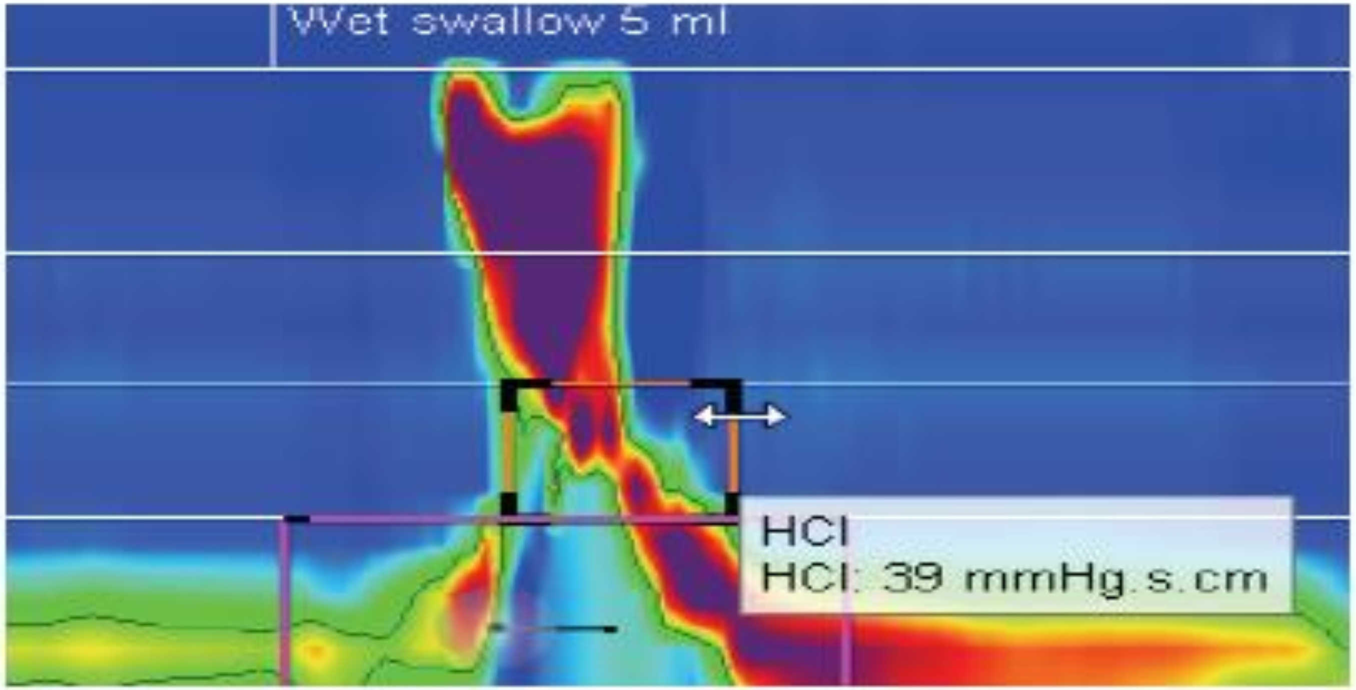
Mean pressure within this domain multiplied by duration (s) and length (cm) In units of mmHg s cm



Hypopharyngeal Contractile Integral (HCI)

Measure of contractile vigor within a space-time box on the pressure topography plot spanning the hypopharyngeal region only

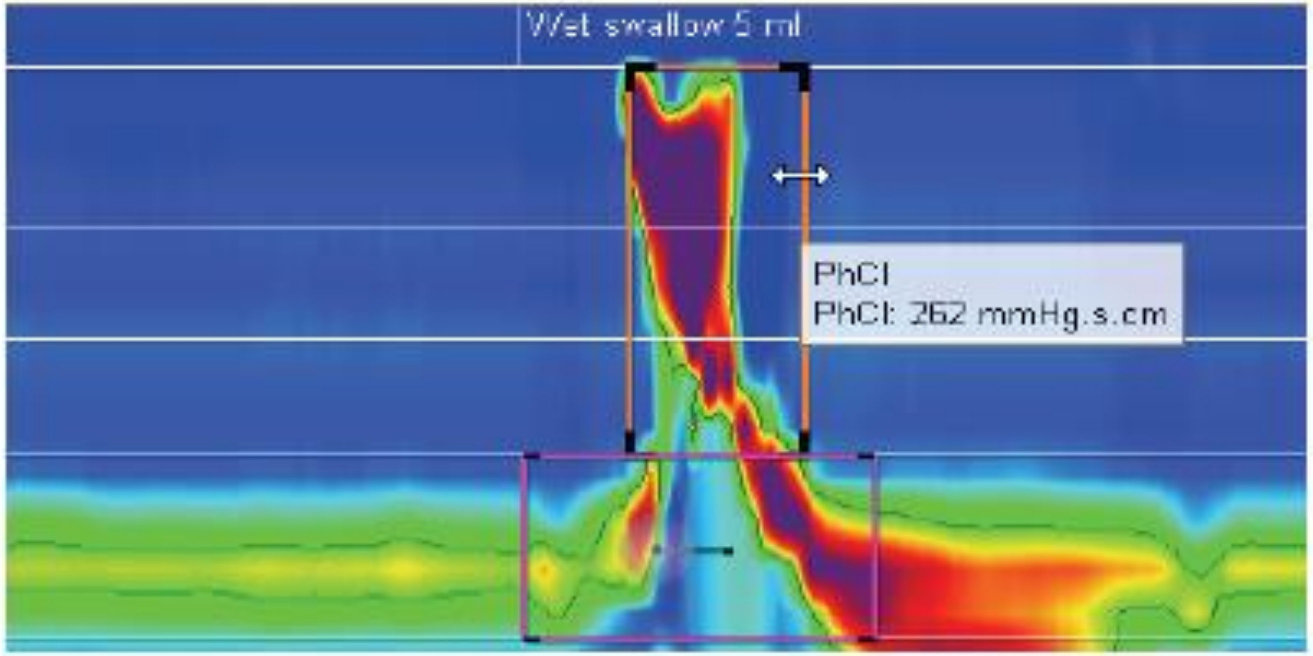
Mean pressure within this domain multiplied by duration (s) and length (cm) In units of mmHg s cm



Pharyngeal Contractile Integral (PhCI)

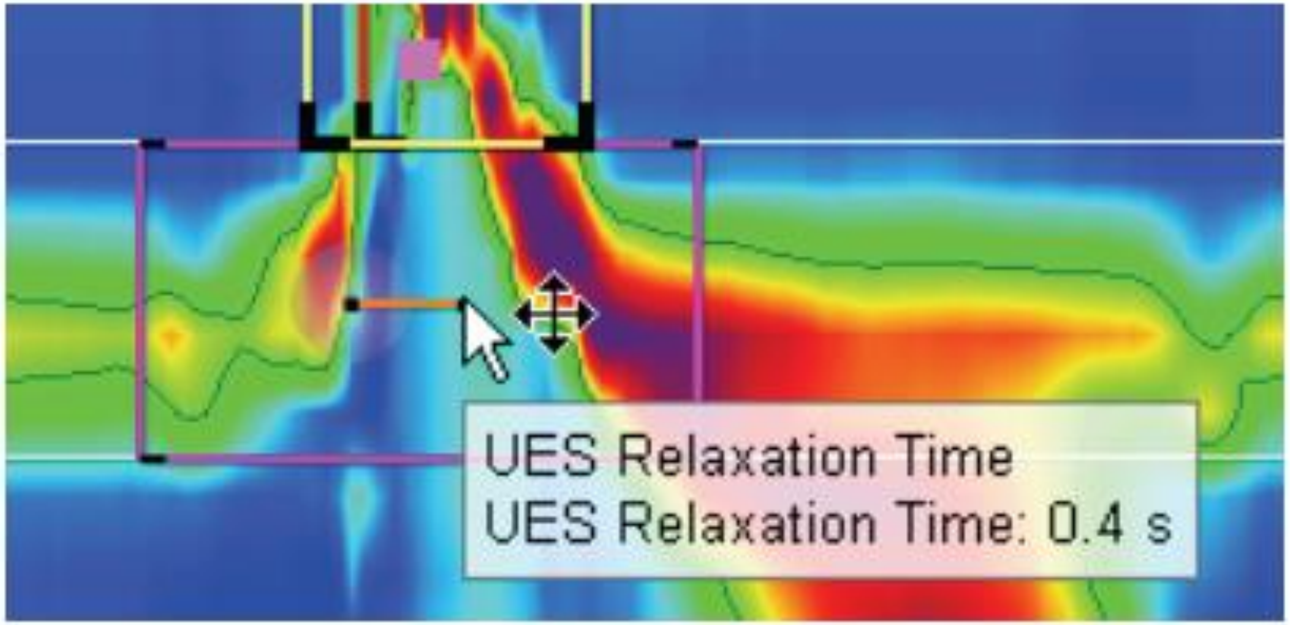
Global measure of the pharyngeal contractile vigor within a space – time box on the pressure topography plot spanning from the velopharygnx superiorly to the upper margin of the UES

Mean pressure withing this domain multiplied by duration (s) and length (cm) in units of mmHg s cm



UES Relaxation Time (UES RT)

Measure of the duration of UES relaxation. It is the pressure interval below 50% baseline, or 35 mmHg, whichever is lower given in units of seconds

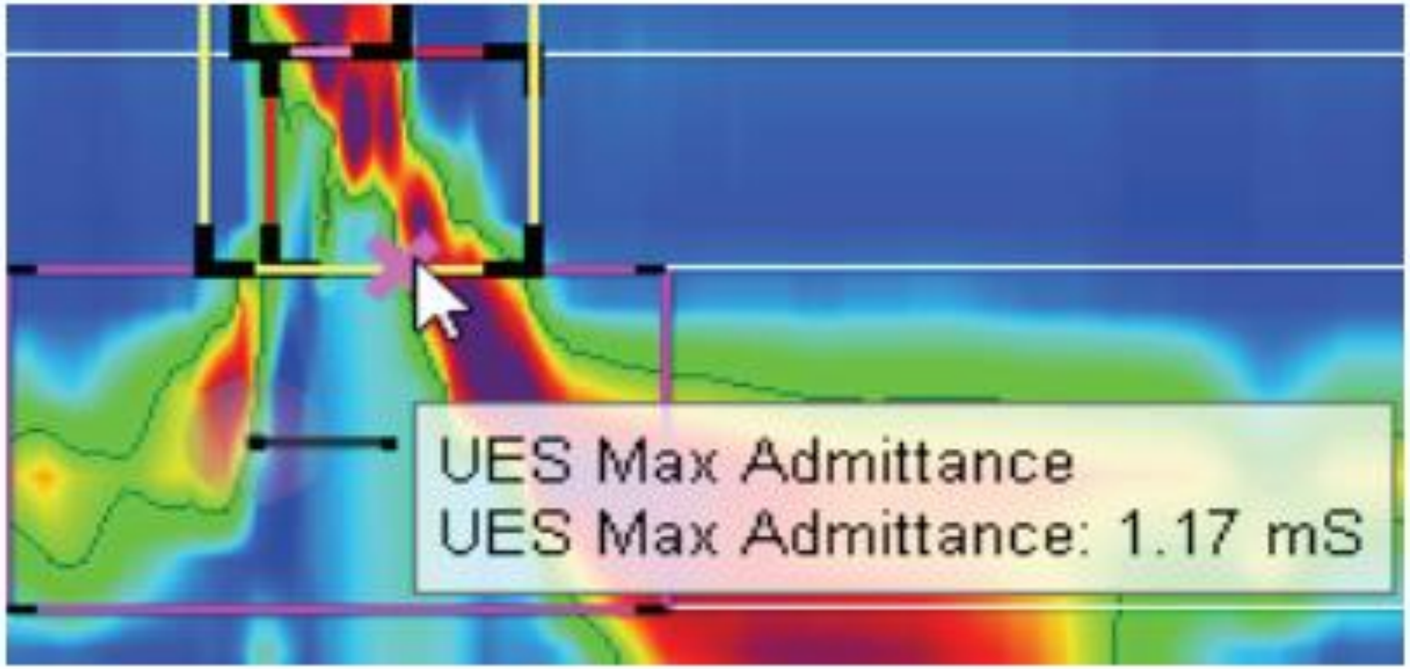


UES Maximum Admittance (UES MaxAd)

Measure of extent of UES opening.
UES MaxAd is the highest admittance value recorded during trans-sphincteric bolus flow in units of millisiemens (mS)

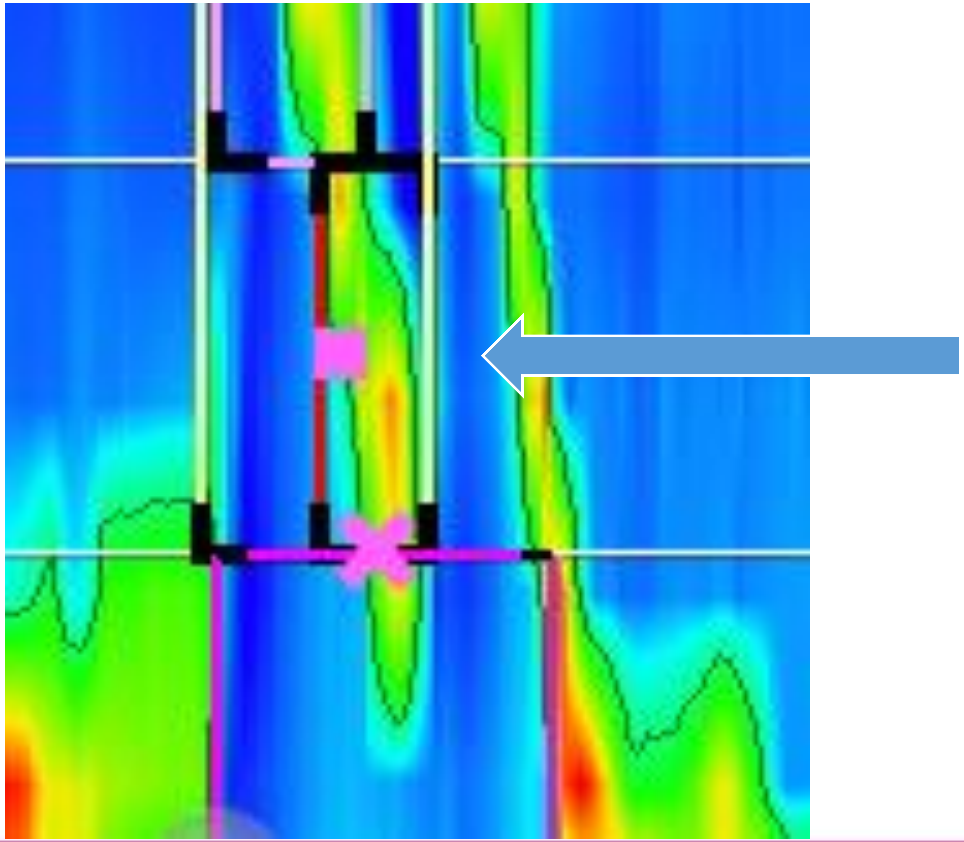
Bolus impedance is a direct correlate of luminal diameter/area. Intraluminal impedance (inverse is admittance) is used as a measure of UES timing and extent of opening

Siemens (symbol: S) is the unit of electric conductance, electric susceptance, and electric admittance in the International System of Units (SI).



Hypopharyngeal Intrabolus Pressure

The pressure 1 cm above the UES apogee position at the time of maximum hypopharyngeal distention deduced from impedance topography in units of mmHg



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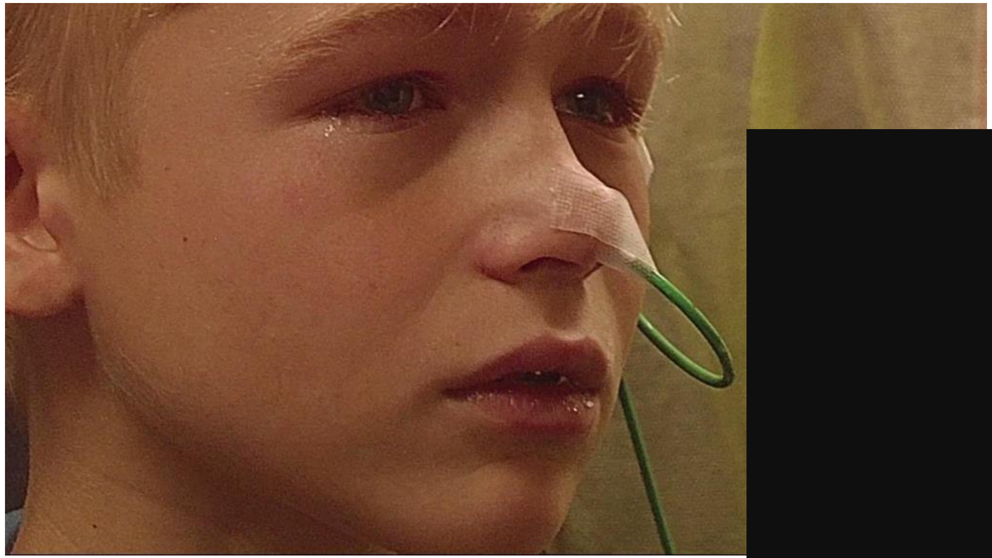
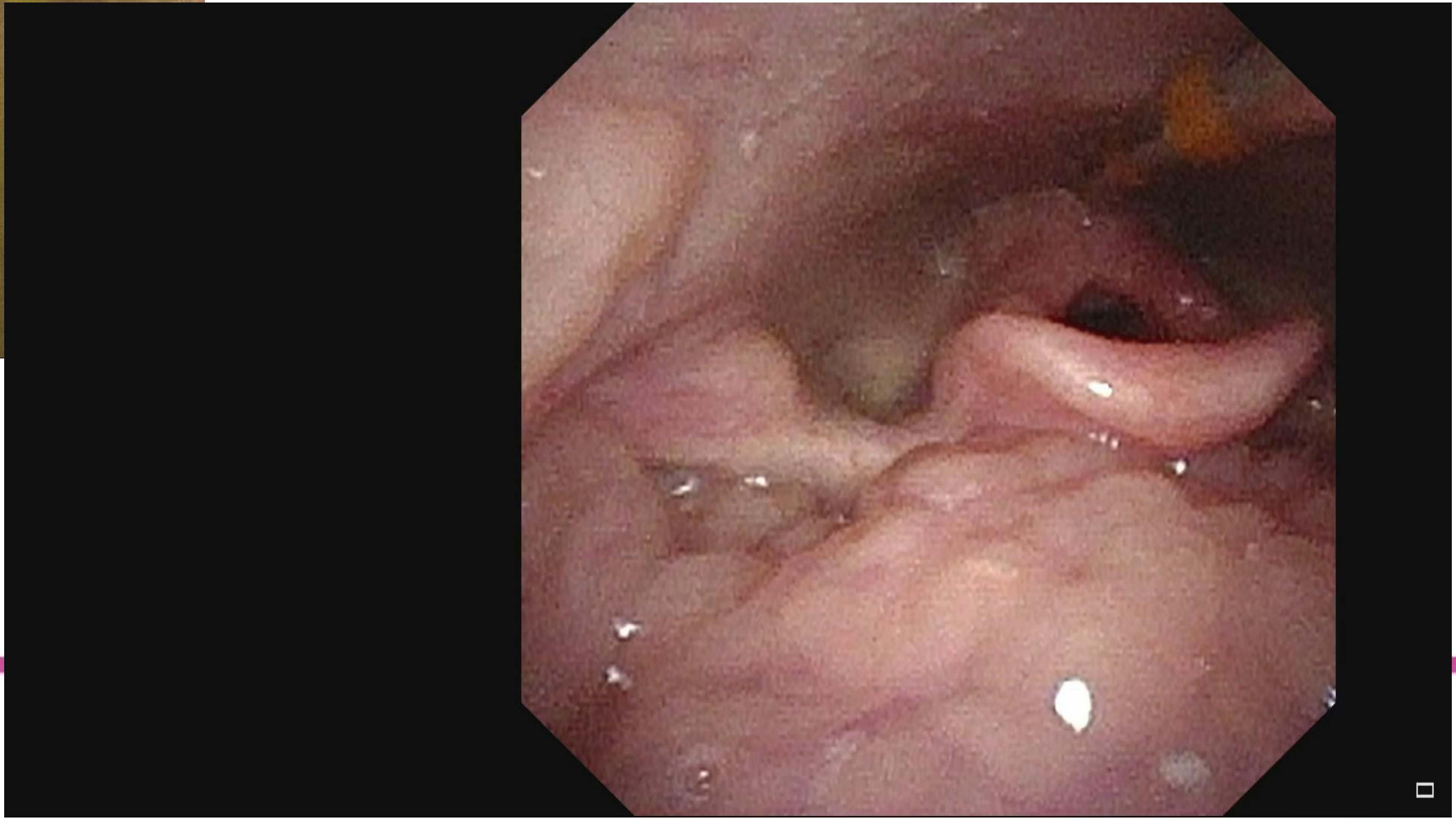
Clinical Presentation Case 1 - TS

- 10 year old male with globus sensation
- Developmentally typical – eating normal diet
- Pre-fees clinical assessment – exaggerated effortful swallow pattern, prior choking episode described

Case Examples: Conditioned Dysphagia





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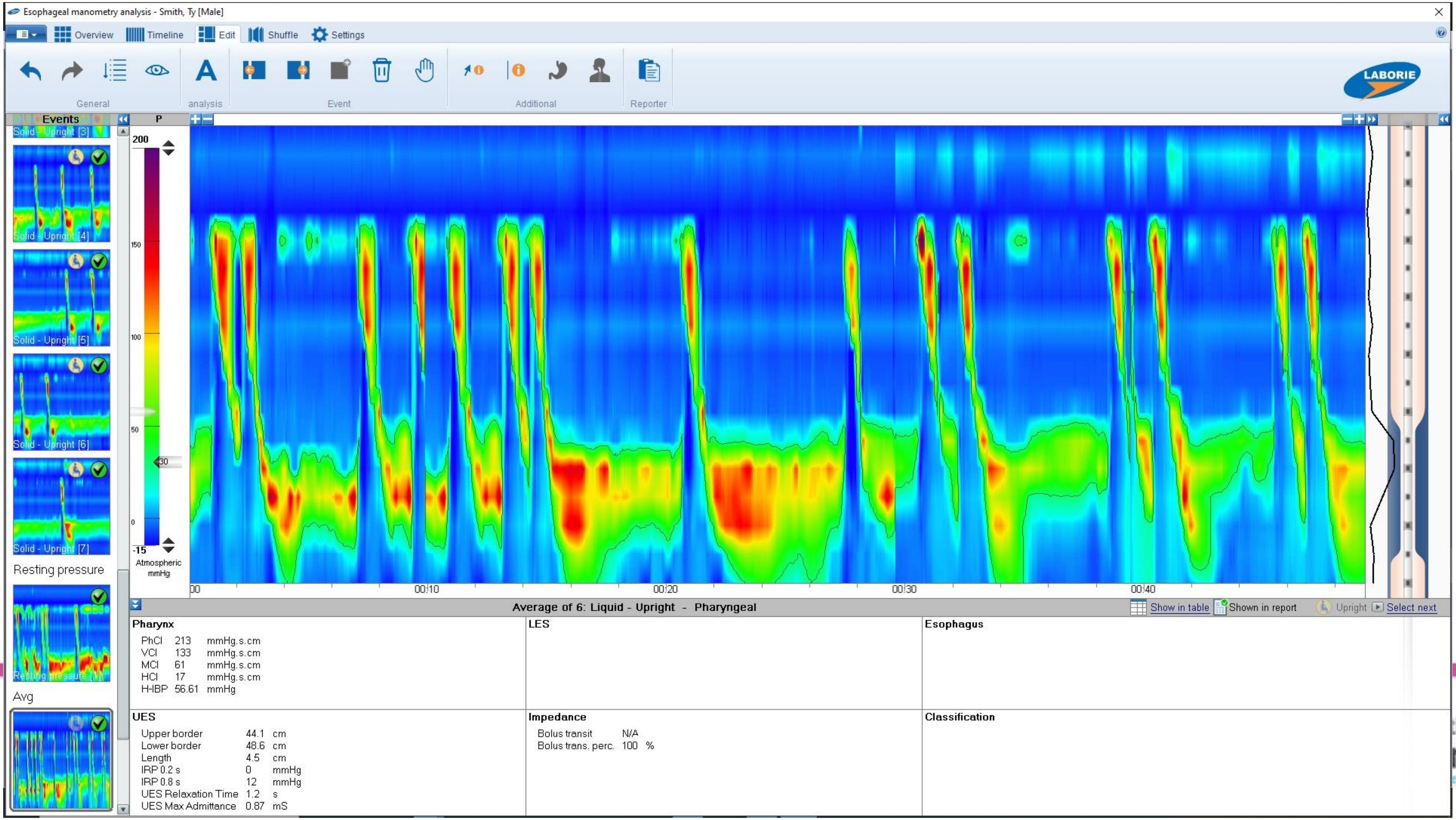
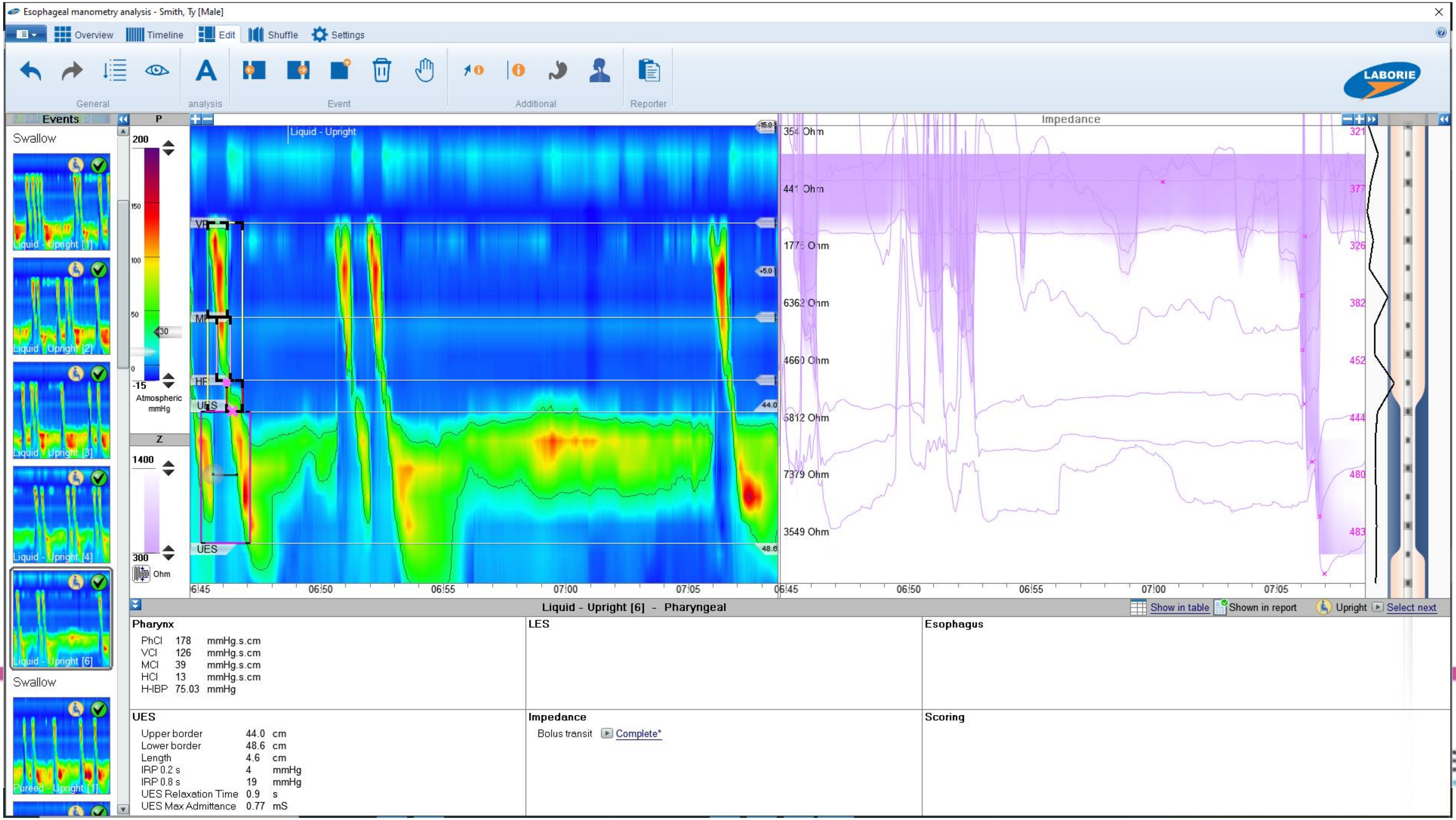


Case Examples: Conditioned Dysphagia



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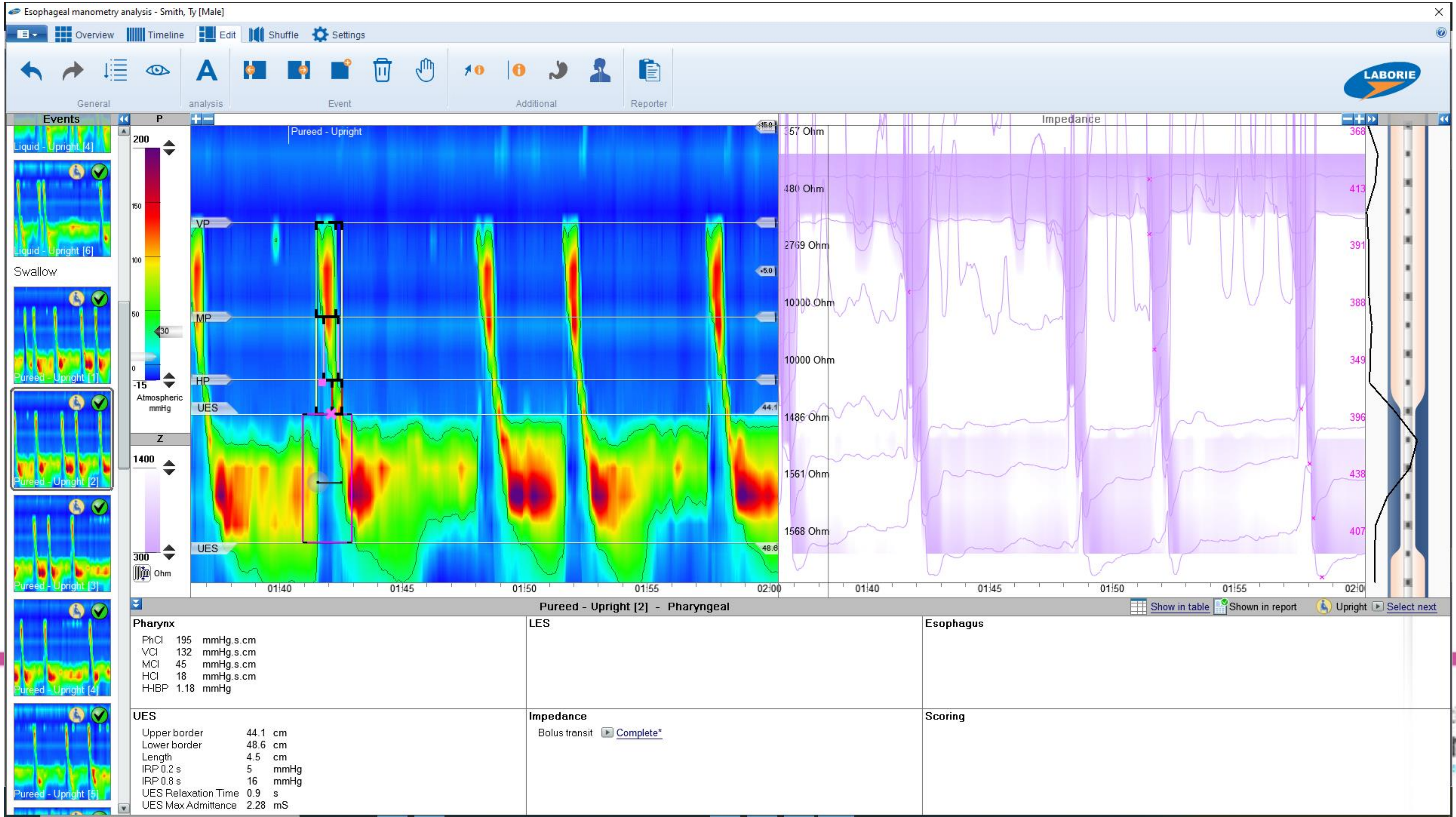
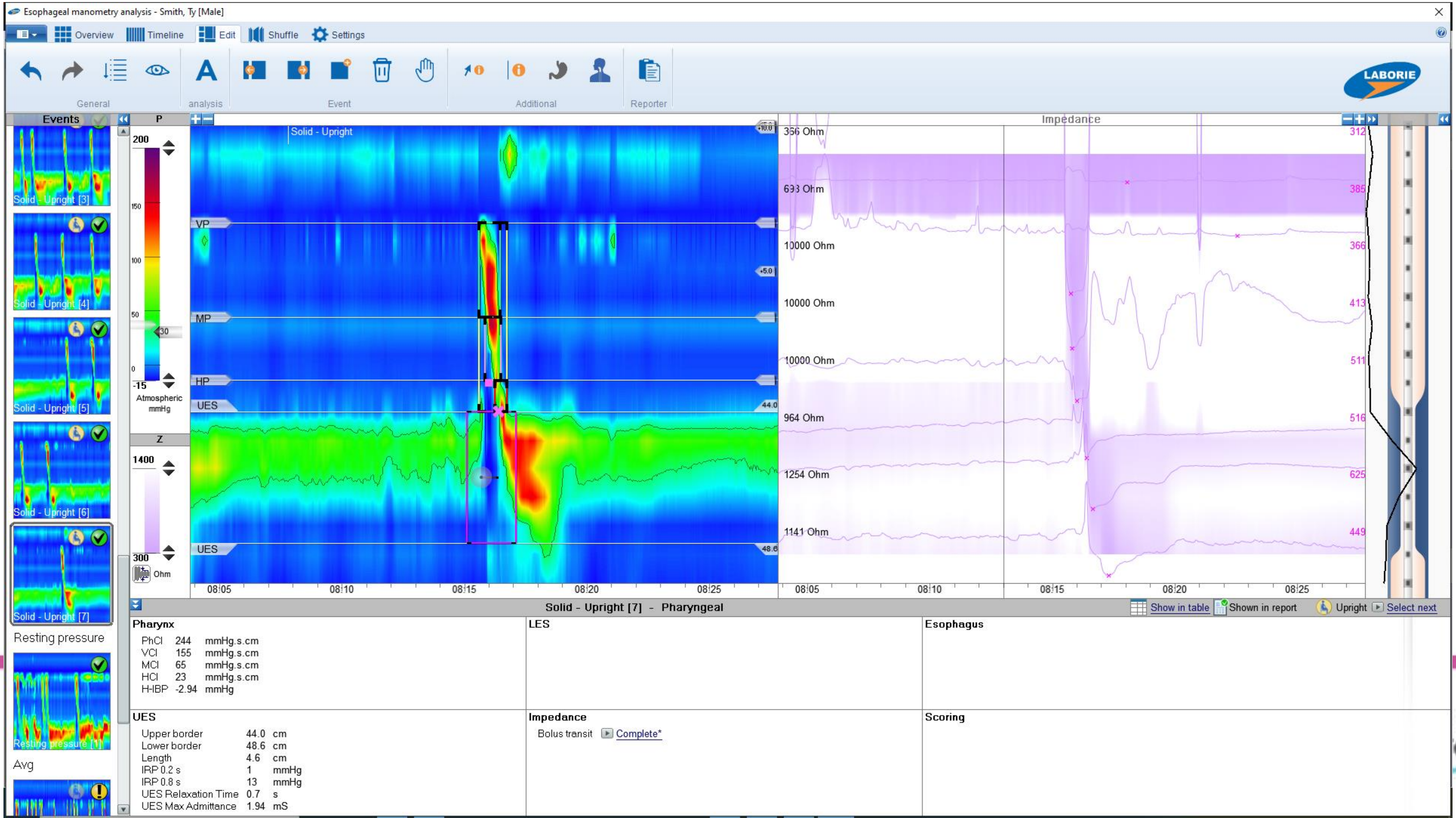


Case Examples: Conditioned Dysphagia



Wrap up Case 1 (TS) – HRPM Findings and Recommendations

- HPRM topography showed typical topography
- FEES results were unremarkable
- Esophagram results unremarkable
- Final recommendation – referral to psychology for treatment

Case 2 HRPM and CHARGE Syndrome

- Genetic syndrome with constellation of clinical features:
  - **C**oloboma
  - **H**eart anomalies
  - **A**tresia of posterior choana
  - **R**etardation of growth and development
  - **G**enital anomalies
  - **E**ar anomalies

  - *CHD7* (chromodomain helicase DNA binding protein) located on 8q12 currently the only gene known to be associated with CHARGE

CHARGE Features

- Variable combination of multiple anomalies
- Varying degrees – *major* and *minor* features
- “Major 4 Cs”= **c**oloboma, **c**hoanal atresia/stenosis, **c**ranial nerve dysfunction, characteristic ear anomalies

Additional CHARGE Features

- Distinctive facial features – square face, prominent forehead prominent nasal bridge, asymmetry from facial palsy
- CNS – cerebral atrophy, corpus collosum agenesis, posterior fossa abnormalities, cerebellar hypoplasia
- Failure to thrive secondary to profound feeding and swallowing issues, motor delay, speech delay
- Cleft lip/palate
- Esophageal atresia/tracheoesophageal fistula
- Endocrine dysfunction
- Immune abnormalities
- Behavioral presentation

Ref: Blake & Prasad, 2006; Hudson et al, *Clinical Genetics,* 2017

***No single feature is universally present or sufficient for the clinical diagnosis of CHARGE syndrome*

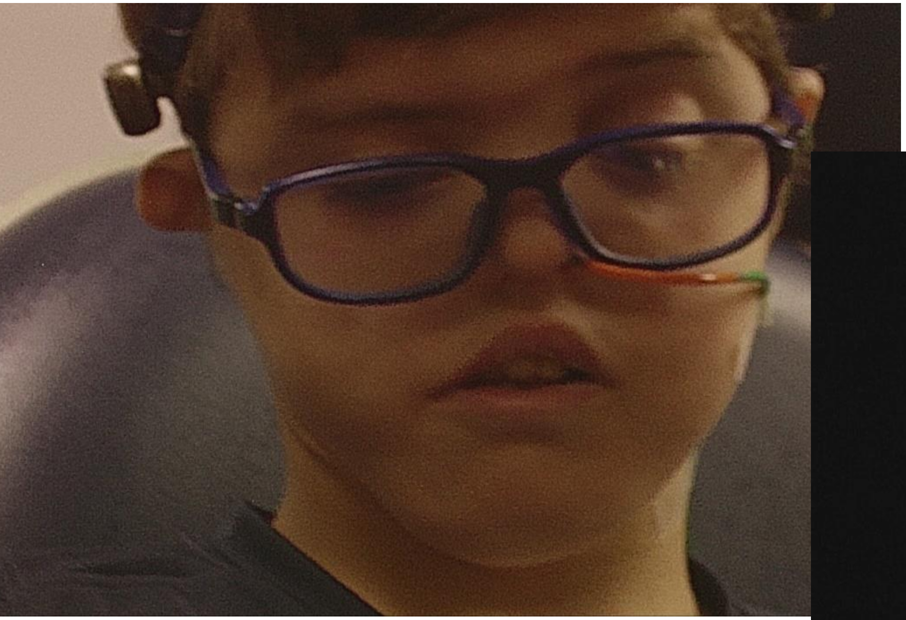
*Cranial Nerve Dysfunction in CHARGE

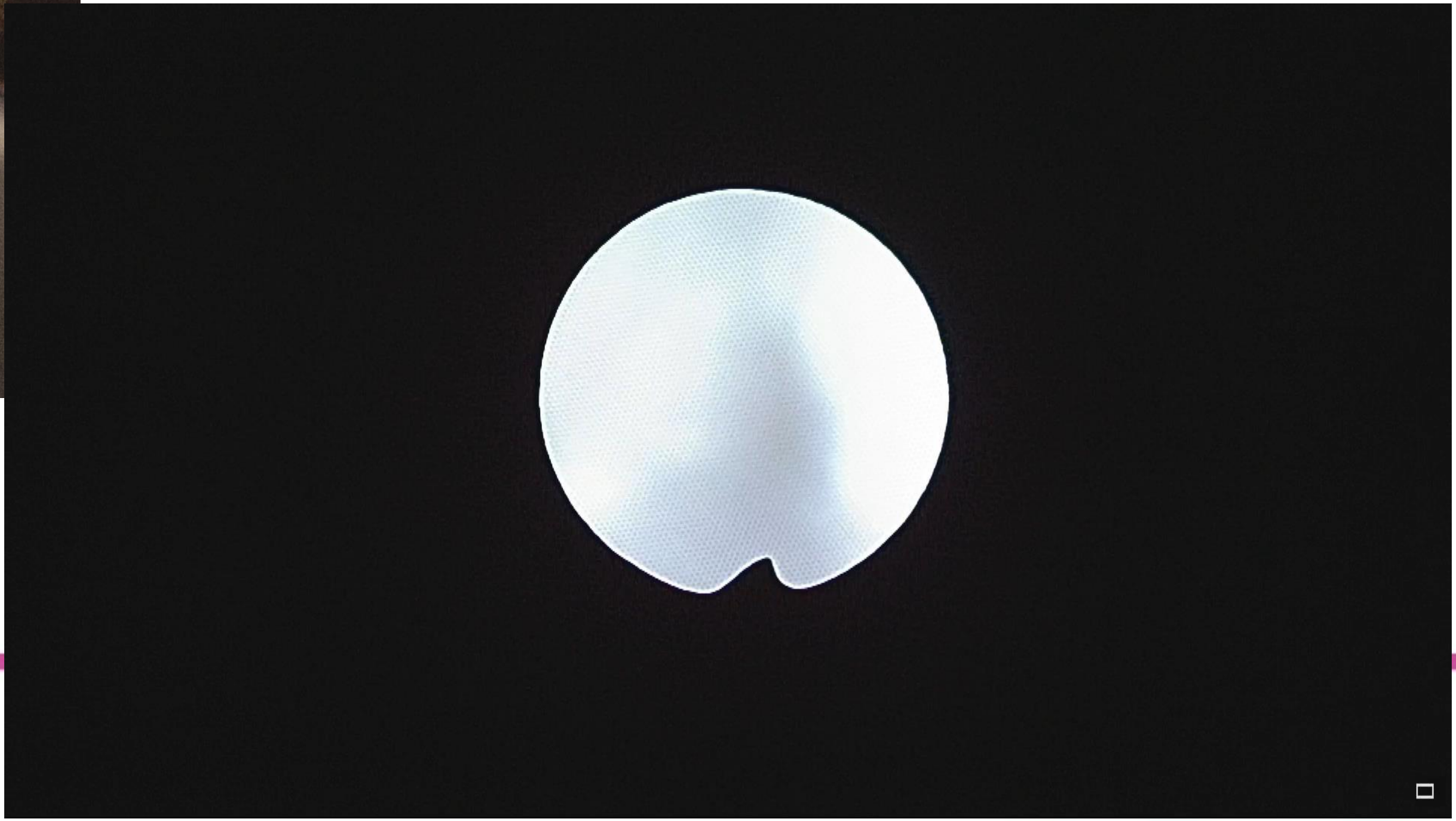
- May be asymmetric
- CN 1 – Absent or malformed olfactory bulb
- CN VII – Facial nerve palsy
- CN VIII – Hearing loss
- CN IX, X, XI – Feeding/swallowing problems, velopharyngeal dysfunction
- CN XII - +/-

Case 2 - CHARGE

- Referred for evaluation to the CCHMC CHARGE Center
- Seeking second opinion regarding overall condition and recommendations for therapy, particularly in regard to swallow function
- Low orofacial tone, poor control of secretions
- Receiving electrical stimulation for swallowing

Case 2: CHARGE Syndrome





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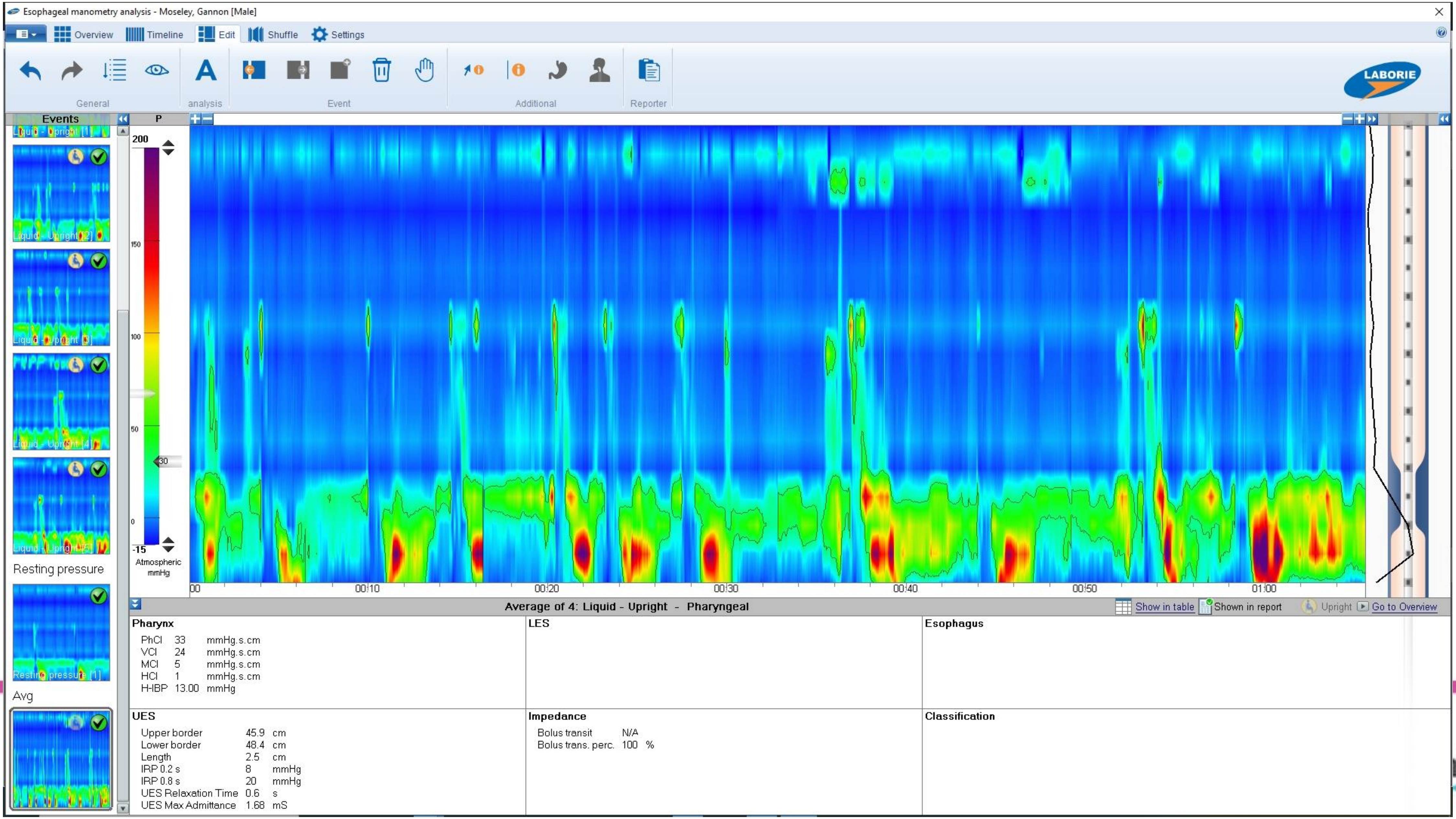

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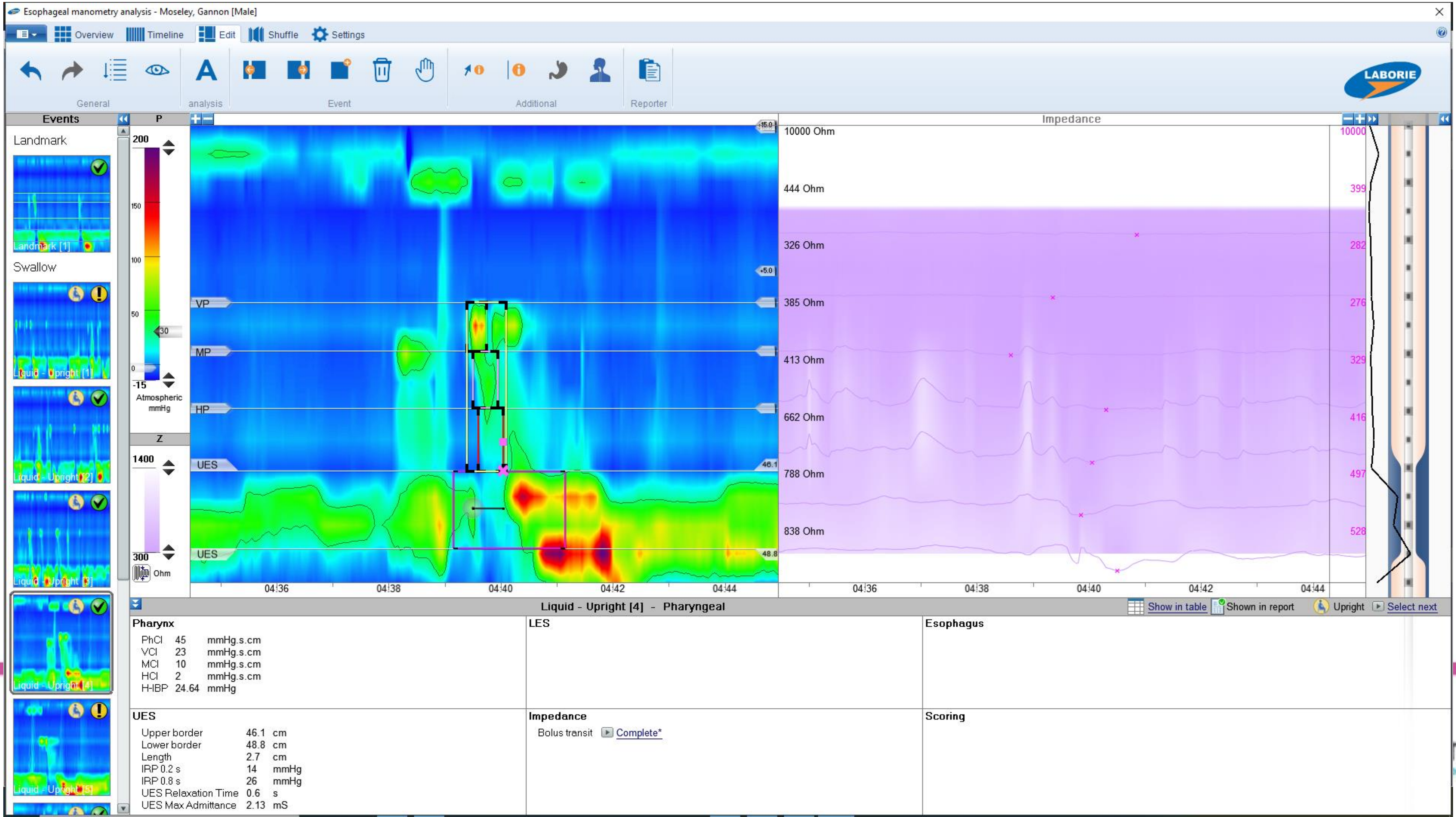
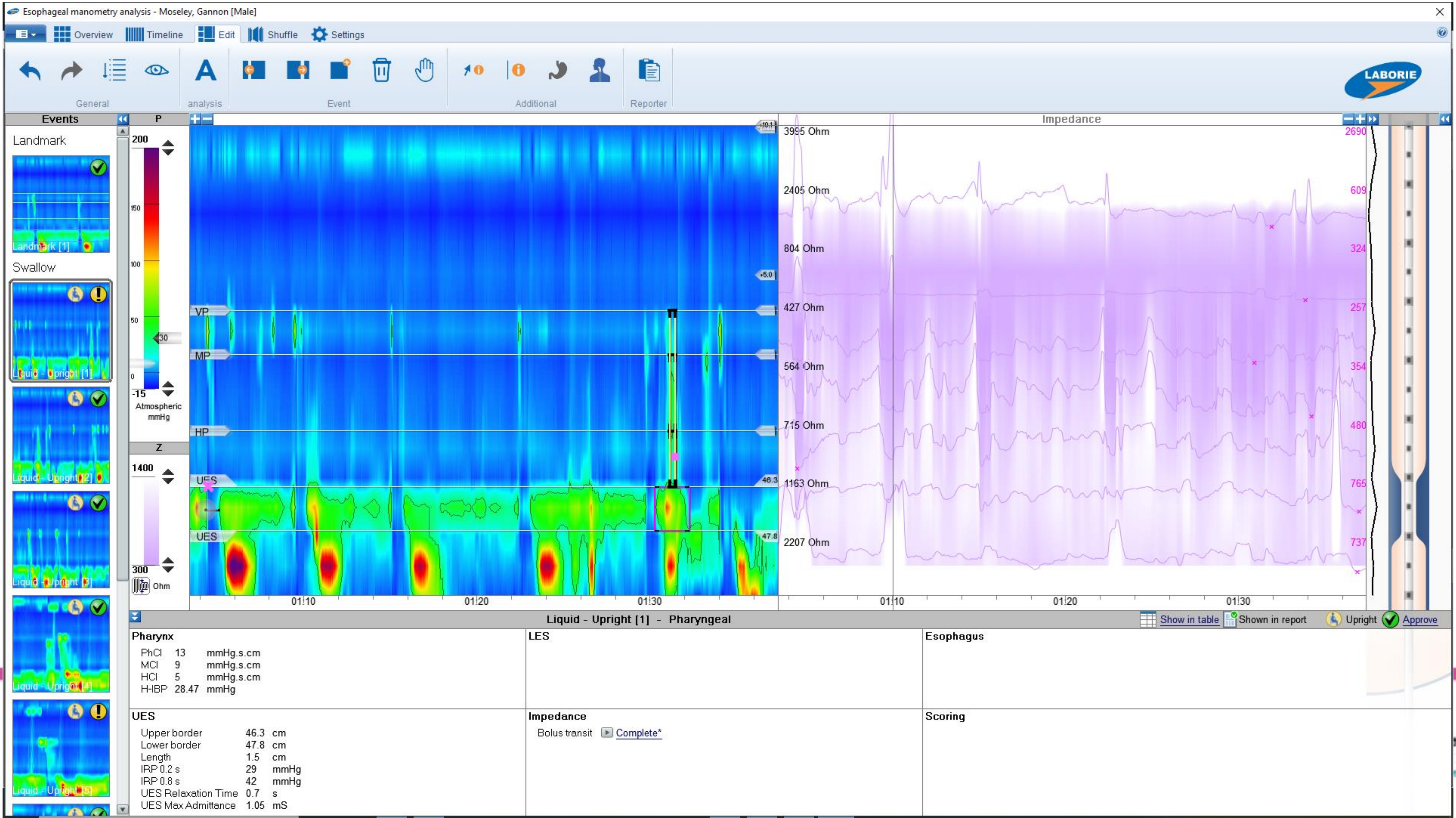


Case Examples: CHARGE Syndrome



Case 2 – CHARGE – HRPM Findings

- HRPM topography indicates inadequate pressure generation
- Implications for treatment recommendations

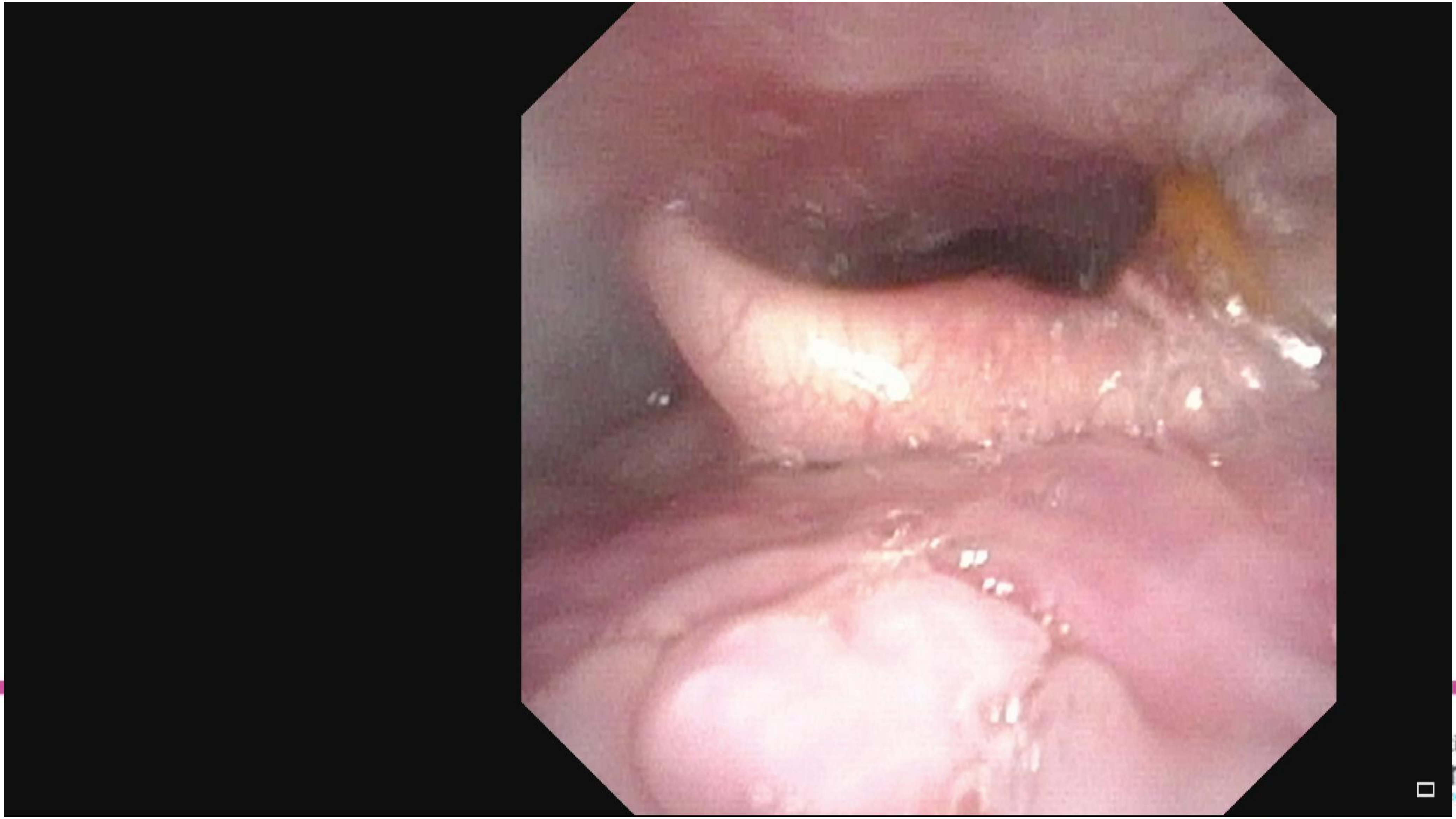
Case 3 – Traumatic Brain Injury

- 12year old male previously healthy
- Motor vehicle accident
- Required cervical fusion – C1-C4
- Spastic quadriplegia, tracheostomy, GT dependent
- Recreational tastes po – with coughing
- Presented for second opinion by Aerodigestive Center of overall status

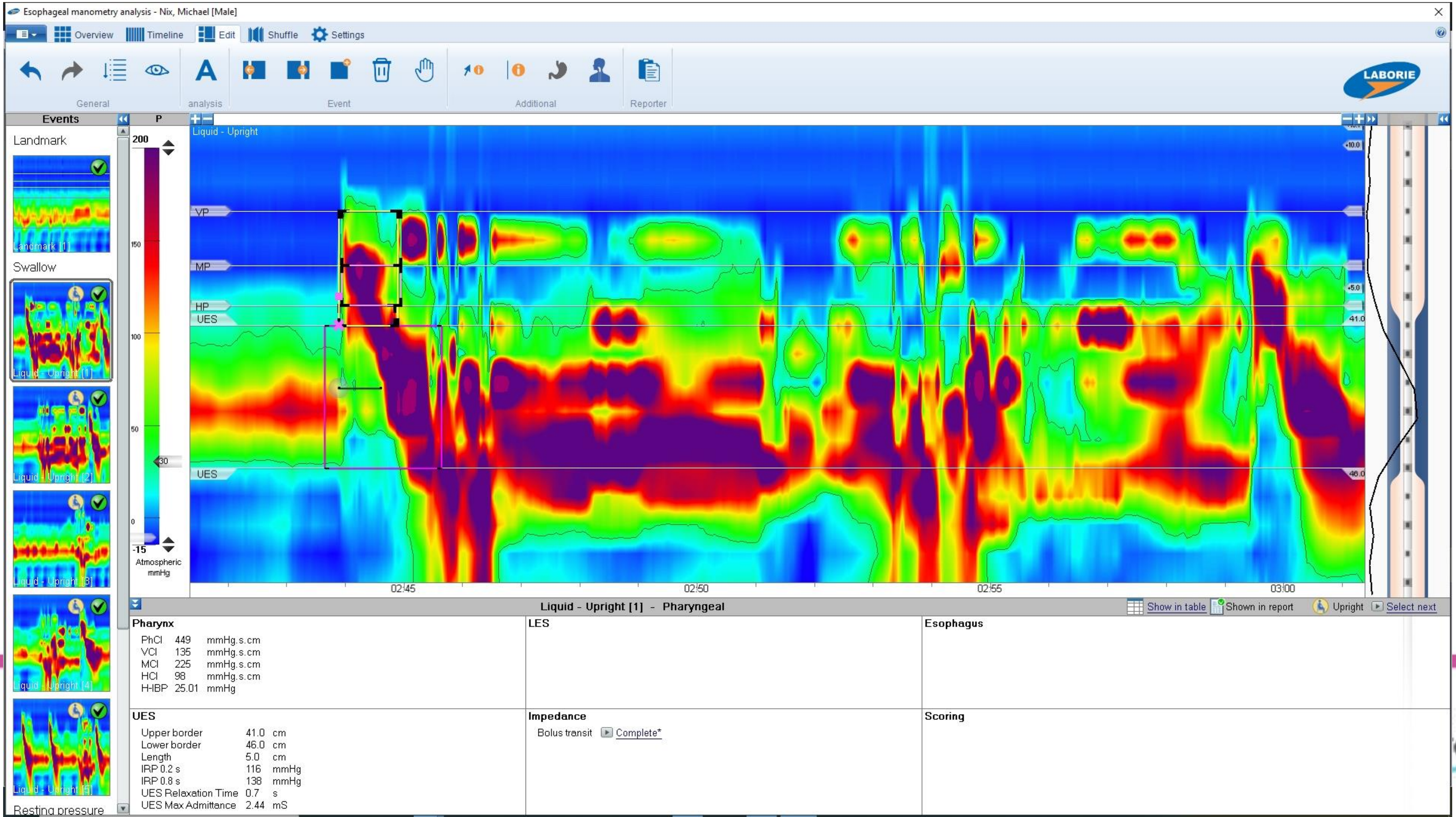
Case 3 - TBI

- Clinical Assessment
  - Limited active oral motor movements – anterior tongue protrusion pattern
  - Glossoptosis
  - Family interested in decannulation and in determining swallowing function

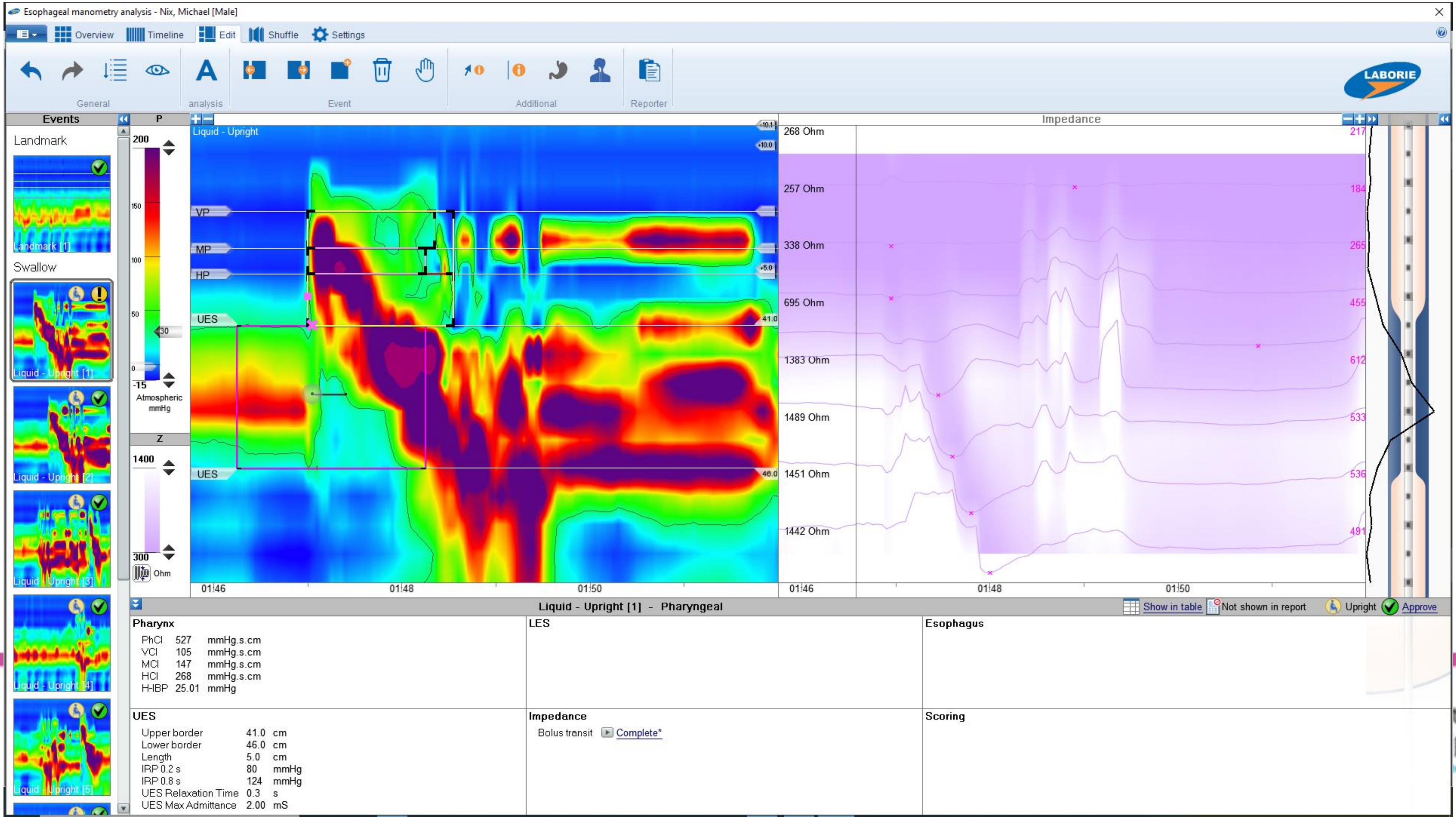
Case Examples: TBI



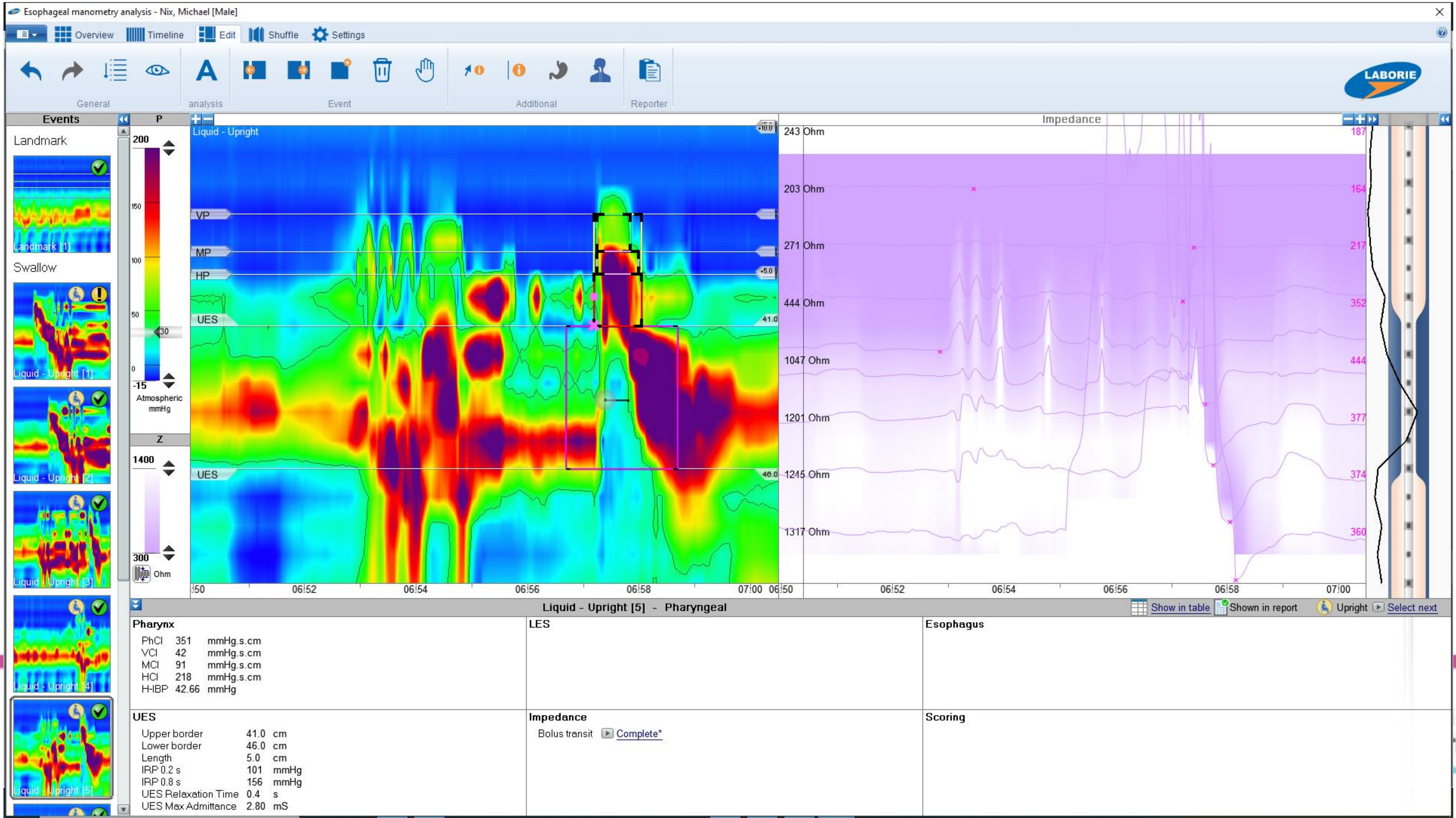
Case Examples: TBI



Case Examples: TBI



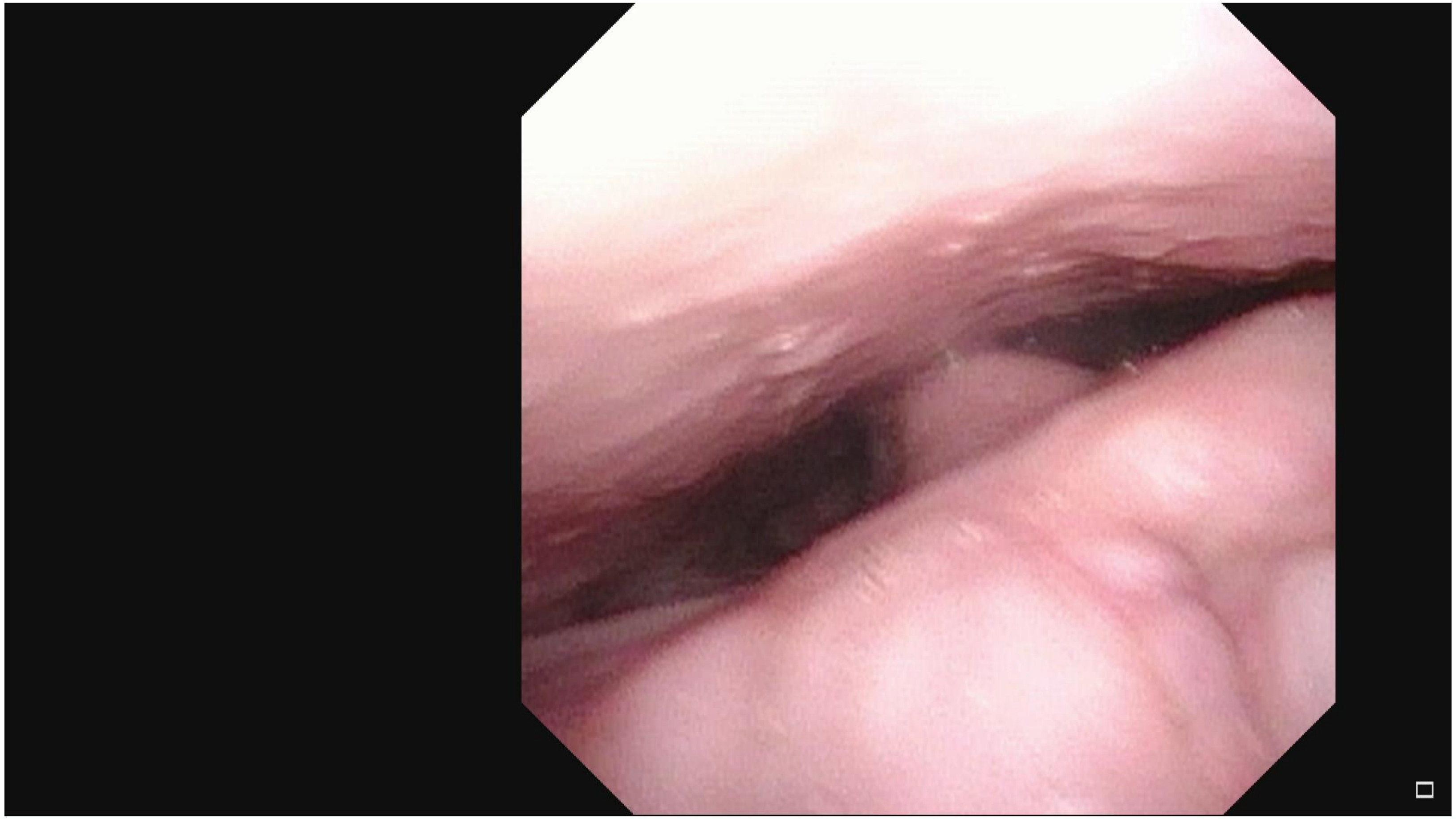
Case Examples: TBI



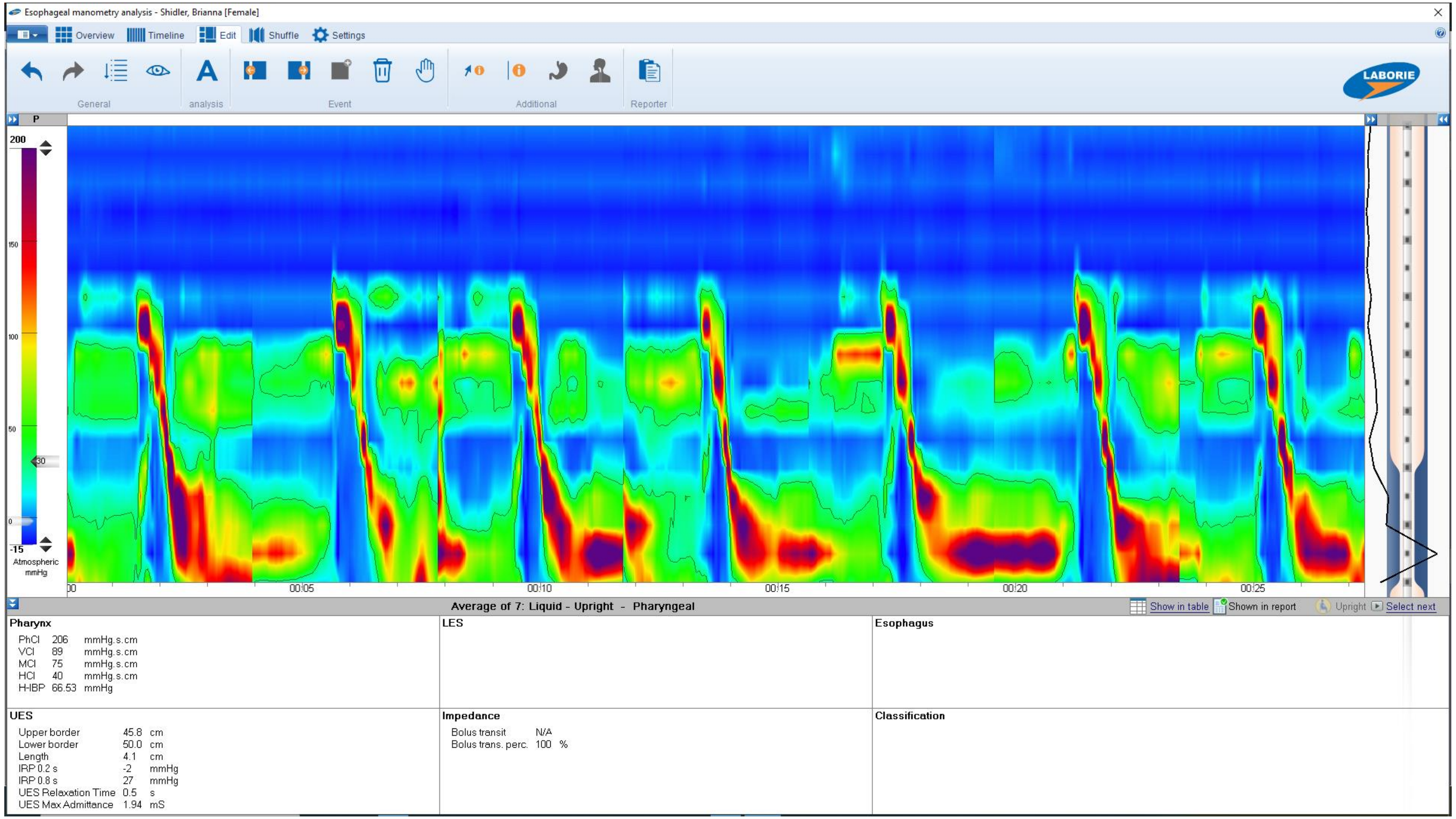
Case 4: B.S.

- 18 year old with Williams Syndrome, Ehlers-Danlos Syndrome (connective tissue disorder – genetic defect in collagen), muscle weakness
- Drinking 62-64 ounces of fluid daily – sudden onset of dysphagia for solids
- Avoidance of solid intake – sensation of sticking, fear of choking

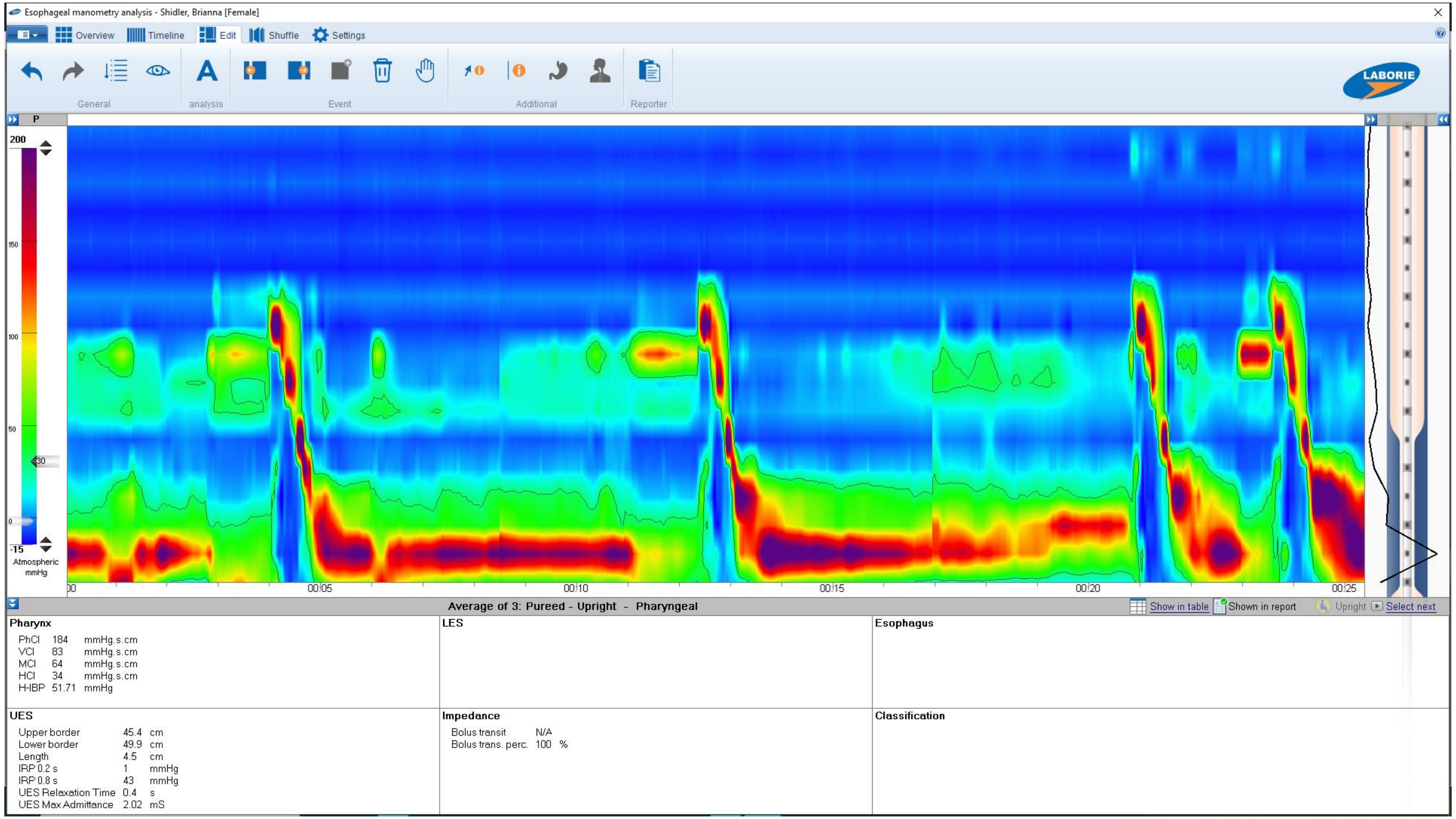
Case B.S.



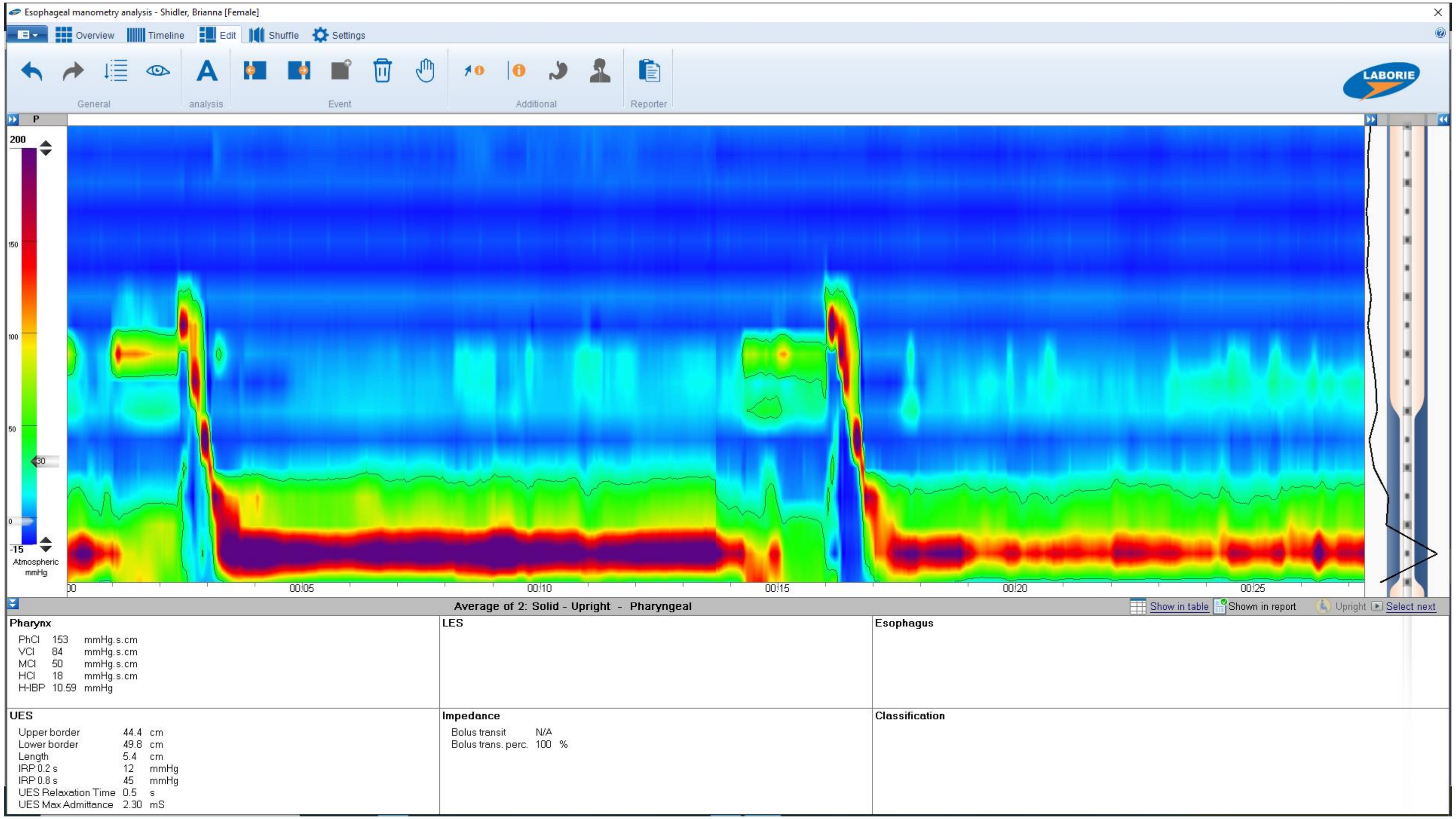
Case B.S. Average Liquid



Case B.S. Average Puree



Case B.S. Average Solid



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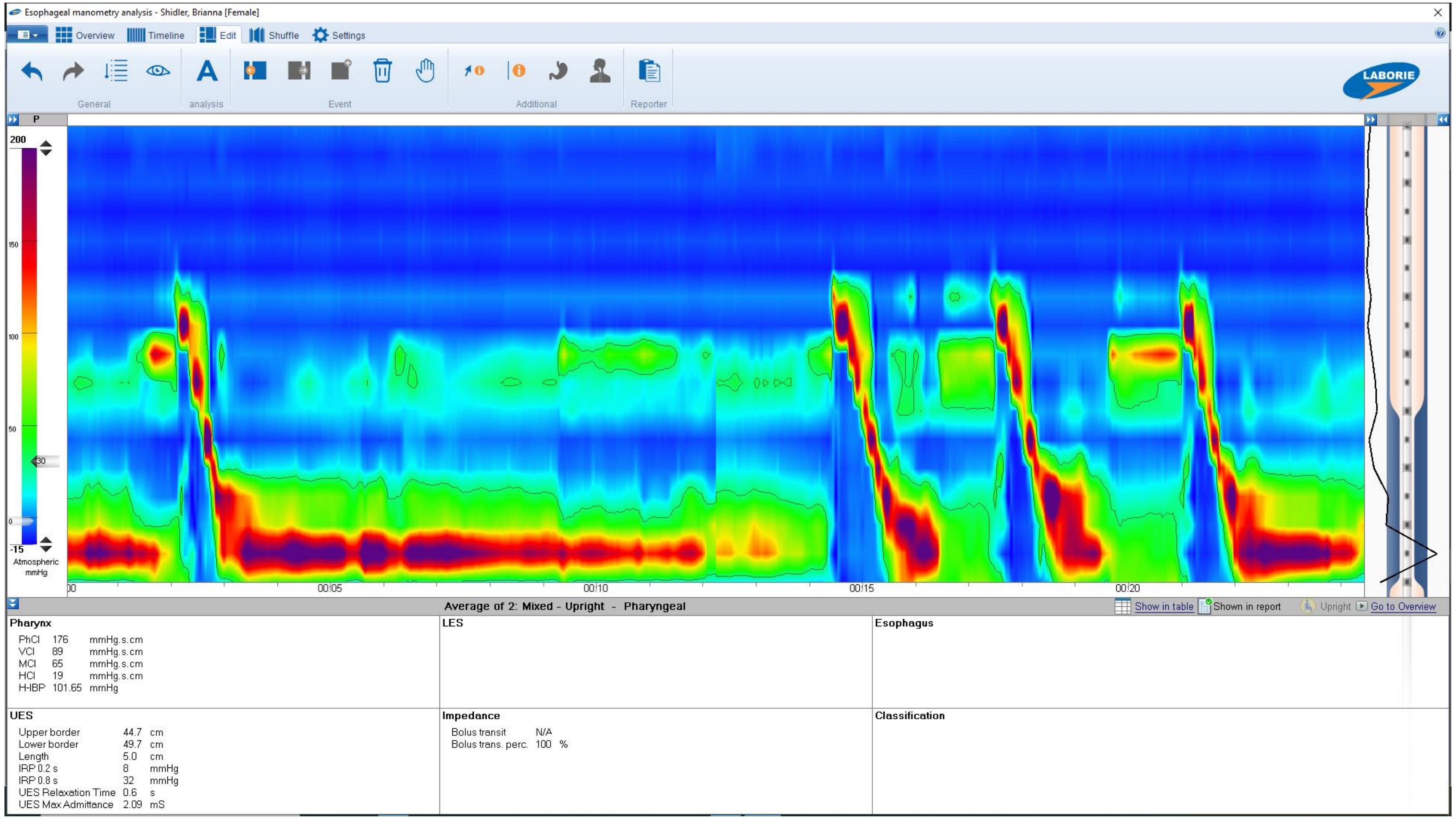
Case B.S. Average Mixed Texture



Questions HRPM Helps to Answer

- Next steps – provides physiologic metrics that may predict type of intervention needed, and data regarding treatment efficacy
- Does this patient have swallowing function – ability to generate pressure during swallowing efforts?
- Helps differentiate specific pharyngeal (weakness), UES (e.g. relaxation vs opening extent), and conditions such as achalasia

Recommend HRPM in Pediatric Patient

- When there is a question of the ability to generate a swallow
  - Syndromic patients
    - CHARGE syndrome
    - Velocardiofacial syndrome
    - Neurofibromatosis
  - Neurologic patients
    - CVA
    - TBI
    - Degenerative disorders
  - Structural abnormalities of the upper aerodigestive tract
    - Congenital anomalies
    - Postsurgical conditions

Limited Value of HRPM in Pediatric Patient

- Children under 1 year of age
- Oral aversion, hypersensitivities
- Limited or no experience with oral nutrition
  - Have not received any therapy for the introduction of food materials
- Chronic emesis problems
- Physiologic condition precludes introduction of oral feeds

Next Steps

- Continue to gather patient data
- Begin to analyze data across ages and conditions
- Compare to normative values for adults
- Help to contribute data to establish normative values for infants and children

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