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Successful evacuation of a PEG probe plate in the oesophagus of a 16 week old child with myotonic dystrophy. An interdisciplinary approach

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Keypoints

This case report shows that the life threating situation of the patient could be overcome with planning (trigger free anaesthesia), good team performance (interdisciplinary, adequate communication) and following the guidelines so that the patient left the hospital without worsening the neurological outcome or general condition.

Abstract

This case report involving an early salvaged patient with a myotonic dystrophy typ 1 outlines the interdisciplinary method to control an unexpected acute life-threatening complication in gastrointestinal procedures in infancy. The only way to guarantee the patient's safety in such a scenario is an exact and interdisciplinary planning and following the guidelines to rescue the foreign body.

Keywords

Airway, respiratory arrest, complications, congenital anomalies and syndromes, pediatric

Introduction

Description of the patient: The concerned patient (former early salvaged in the 30 4/7 pregnancy week), 16 weeks old, suffers from a myotonic dystrophy type 1. Myotonic dystrophy type 1 is a form of myotonic muscle disease with muscle dystrophy. It is the most common muscle dystrophy with an incidence of 5/100,000 a year. The inheritance is autosomal dominant. It is a matter of a defect on the chromosome 19 with a (CTG)n Trinukleotid-repeat-expansion (unstable CTGrRepeats in the DMPK-gen). The cause of this defect is the reduced production of the myotonin-protein kinase and through that a muscle fibres and calcium pump SERCA defect. It exists in a congenital form with a manifestation at birth as a floppy infant and an adult form (in the second or third live decadent). The adult form presents itself with progressive muscle weakness especially in the face and legs, heart rhythm disturbances, and cataract. The examination and diagnosis took place with measuring the electrical muscle activity (EMG) and direct gendiagnosis out of leucocytes. The disease is not curable. The life expectancies are in the average range of 50 -60.

After birth this patient had to be admitted because of a respiratory global insufficiency with lack of surfactant and insufficient breathing muscle. During the period of treatment at the intensive care unit, the patient showed a respiratory improvement. The hard case of myotonic dystrophy manifested with the floppy infant and drinking weakness. The initial kidney failure showed improvement during the stay. The gastroenterological treatment concept is the therapy of the trouble because of the dysphagia with a PEG probe. An improvement of the symptoms was strived for with physiotherapy, ergo therapy, and speech therapy. As part of the genetic investigation a mild form of the syndrome (MDT1) was diagnosed in

the relatives on the mother's side. The mother of the patient claimed a light form of the muscle weakness; the grandmother did not show any symptoms.

The goal of the anaesthesiology is the prevention of any myotonic reactions with a rise of potassium or rhabdomyolysis. The use of depolarisating muscle relaxation as well as volatile anaesthesia is prohibited because of the danger of a malignant hyperthermia. Because of that we delivered an intravenous anaesthesia with propofol, benzodiazepine, and opioids. The desire was to have sufficient muscle strength so that the patient could breathe spontaneously without pulmonic complications. Non-depolarisatic muscle relaxation is used intermittently; cholinesterase inhibitors are prohibited because of a depolarisation block. (1)

Description of the intervention: A tube was inserted through the skin into the stomach directly during the procedure of a percutaneous endoscopic gastrostomy. The "pull-through-method" is described as a procedure where a needle is inserted with localisation of the puncture spot through the stomach wall from the inside. Then a string is inserted through the needle into the stomach and the PEG probe plate is pulled from inside to outside. After pulling the probe, the plate remains in the stomach as an abutment. The gastric wall is pulled and fixed at the abdominal wall. (2)

Case report

Case representation: Primarily all necessary preparatory measures for a trigger free anesthesia were prepared: cleaning the system, changing the hoses, and vapor expansion. The anesthesia was performed intravenously with Propofol and Remifentanyl. No relaxation was used. Initially it was difficult to put an IV into the patient and no inhalation anesthesia was attempted because of the muscle disease. A cuffed tube was inserted into the trachea. The reflection of the esophagus and stomach to lay the string was, at first, complication-free. Afterwards the incision was induced. During insertion of the PEG-plate no problems accrued at the upper part of the esophagus. Then a resistance was felt so that the PEG-plate was not movable. The breathing pressure rose to 25mbar, the capnography disappeared, and the oxygen saturation dropped to 80%. Even though we ventilated manually with pressure over 30mbar, there was no improvement. We interpreted this situation as an obstruction through the probe in the esophagus to the trachea. As a differential diagnosis it could have been a pneumothorax, mediastinal emphysema with compartment syndrome, gastral perforation etc. A 18 CH peripheral venous catheter was inserted into the stomach to release the air in the stomach because of the distended abdomen and the theory that that abdomen was pushing into the thorax. No air came out. Another gastroscopy showed a lesion (1/4 circumference) in the upper esophageal part with a via falsa. This caused a thoraxemphysem. The oxygen saturation dropped to 55%, at the same time no carbondiaxoid was measured. As a differential diagnosis a pneumothorax on both sides was suspected so we punctured both lungs, but again no air came out. The patient had a bradycardia of 60 bpm and was resisted for about 180 seconds. At the same time he received 1mcg adrenaline. After stabilization he received a suprarenin-perfusor with a dose of 1 – 2 mcg/min. To investigate the cause of the pulmonic situation a thoracic x-ray was taken. It showed mediastinal emphysema, but no pneumothorax. After improvement of the cardio-pulmonic situation a procedure was thought over. Plan A was to push the plate back into the stomach, plan B was to remove it with the gastroscopy orally. A couple of frustane attemps to secure the plate (caudal or cranial) followed. A thoracotomy to recover was planned. For that the patient had to be turned on the left side to approach the esophagus. After a short interdisciplinary debriefing about the situation the plate was recovered with a videolaryngoscopy and Magill-forceps. If that had not worked out the other plan would be to leave the probe plate in the esophagus and to stabilize the patient on the ICU to do the removal later. Under stable cardiopulmonary conditions a feeding tube was inserted

endoscopically and an endosponge was inserted into the lesion. The endosponge helps heal the wound by sucking the sore secretions. To guarantee enteral nutrition, an open gastrostomy was established in the left upper abdomen. After finishing the intervention the patient was transported into the intensive care unit. After 24 hours the patient was stable and showed an inconspicuous EEG. Four days afterwards another intervention followed where the tracheal tube was changed from oral to nasal and a change of the endosponge was accomplished. 2mg Rocuronium and 15mcg Fentanyl was applied. Seven days after the first intervention a healing of the lesion was shown. Again the patient received only 1mg Rocuronium and 30mg of Propofol. On the ICU the patient was extubated and on the ninth postoperative day and shortly thereafter the patient was transferred to the normal ward. Two weeks later the gastroscopy showed a total healing of the esophageal lesion. The patient was released home in good general and nutritional condition (Figures 1,2,3)

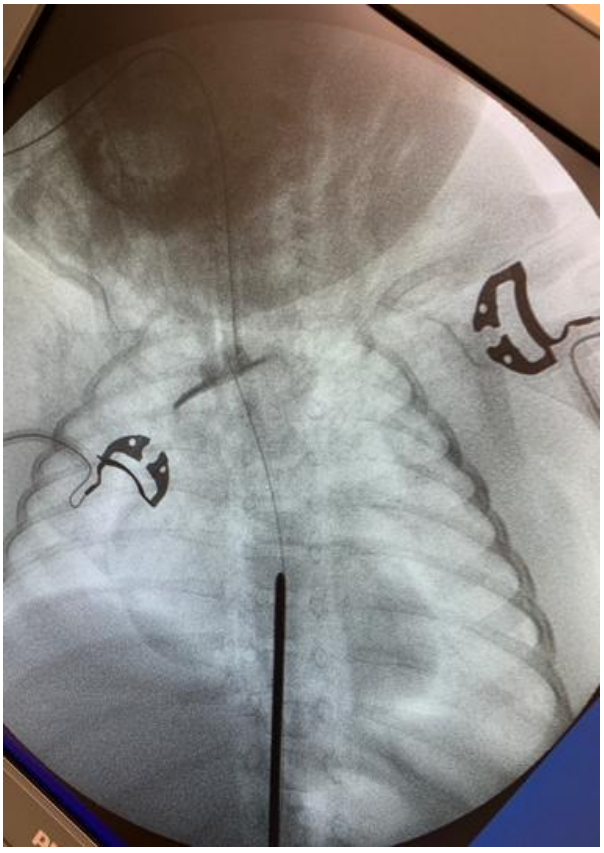

Figure 1 Thoracic X-ray of the patient with the stucked probe plate in the esophagus

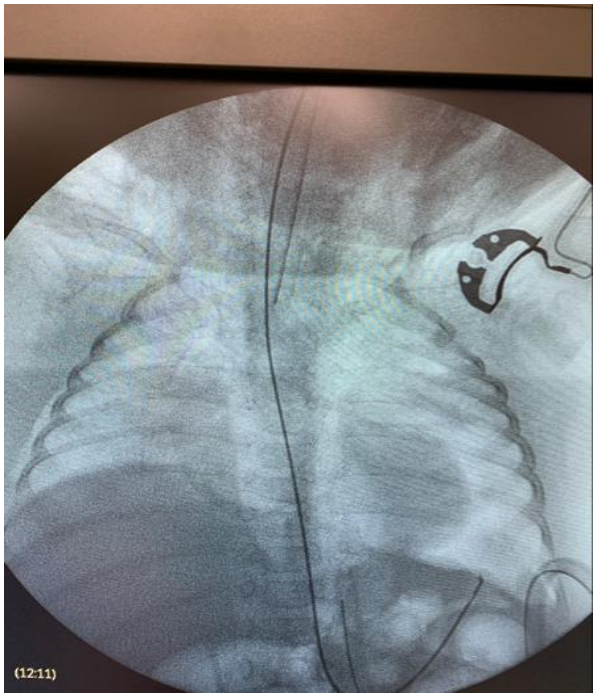

Figure 2. Thoracic X-ray of the patient immediately after rescue of the probe.

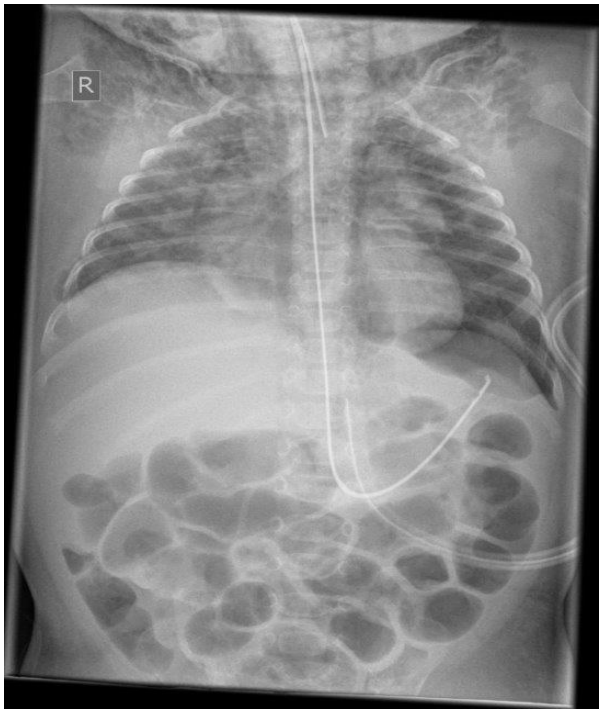

Figure 3. Thoracic X-ray of the patient with skin emphysema after the rescue of the probe plate and the insertion of the feeding tube

Discussion

This case report outlines the interdisciplinary method to control an unexpected acute life-threatening complication in gastrointestinal procedures in infancy. We orientated ourselves with the S2K-Guidelines of foreign body in children of the year 2015. They

recommend the use of videolaryngoscopy (benefit – better view, teamwork and documentation) or laryngoscopy by foreign body aspiration. To remove the foreign bodies, use a barrel instrument like the Magill-forceps. If you have foreign body ingestion (like in this case) one uses the videolaryngoscopy or laryngoscopy to have a good view on the hypopharynx. If it is possible to visualize the foreign body one should extract the foreign body with forceps. Foreign bodies which lay deeper in the esophagus should be removed with a flexible or rigid esophaguscopy or gastroscopy (Kay 2005, Russell 2014). Both flexible and rigid should be available. Depending on the type of location of the foreign body one may have to push it back into the stomach to establish an intestinal passage. This push forward requires awareness and sensibility. (3) In our case the foreign body was located at the height of the clavicula respectively thoracic vertebral body one. That was the reason why we could remove it with the Magill forceps and videolaryngoscopy. In our hospital it is a requirement to intubate the newborn with a videolaryngoscope because of its benefits (better view, teaching, four eyes principle etc.). (4) A therapy with endosponge by esophageal lesions is more established for adults than for children. (5) Because of multidisciplinary work heavy complications could be solved. Good communication is very important to overcome stressful situations and to find strategies. The Crew Resource Management helps us to optimize. (6) It showed that 70 percent of the human factors (situation awareness, fixation mistake etc.) are avoidable and that you need CRM to overcome these. (7) To criticize us video- or photo documentation on the videolaryngoscope should be required. That is the reason why we used our cell phone camera to document.

Conclusion

This case report shows that the life threating situation of the patient could be overcome with planning (trigger free anaesthesia), good team performance (interdisciplinary, adequate communication) and following the guidelines so that the patient left the hospital without worsening the neurological outcome or general condition. The only way to guarantee the patient's safety in such a scenario is an exact and interdisiplinary planning.

Diagram:

PEG-probe not movable in the esophagus
↓
Co2↓, ventilationspressure↑, SpO2↓
↓
abdomen distended, DD: air in the abdomen, pneumothorax
↓
incision with needle → no improvement
↓
gastroscopy →lesion, DD: thoraxemphysemia, 55% spo2
↓
incision of the lung
↓
Bradycardia 60 bpm
↓
resuscitation for 180 sec. + 1mcg adrenalin
↓
stabilisation with 1 -2 mcg suprarenin
↓
diagnostic → thorax x-ray → thoracic emphysema
↓
Frustane attamps to secure the probe (caudal as well as cranial)→ plan of recovery with a thoracotomy
↓
interdisciplinary debriefing
↓
videolaryngoskopy + magill forceps → thoracotomy
↓
recovery of the probe

Plan: A. pushing the probe into the stomach
B. removing the probe out of the esophagus
C. removing the probe via videolaryngoskopy plus magill forceps
D. thoracotomy
E. leaving the probe in position plus stabilizing the patient on the ICU to remove it later on

References

1. Biro P., Vagts D., Emmig U., Pasch Th., Anästhesie bei seltenen Erkrankungen, published 2005, Springer-Verlag Berlin Heidelberg
2. Eich C. B., S2k-Leitlinie Fremdkörper Kinder 2015, Arbeitsgemeinschaft der Wissenschaftlichen Medizinischen Fachgesellschaften e.V
3. Jagannathan N., Sohn L., Hajduk J., Troubleshooting Technical Difficulties With Videolaryngoscope Use in Children: Initial Steps Toward Improving Tracheal Tube Passage, Anesth Analg 2018 ;127:340-341
4. Still S., Mencio M., Ontiveros E., Burdick J., Leeds SG., Primary and Rescue Endoluminal Vacuum Therapy in the Management of Esophageal Perforations and Leaks, Ann Thorac Cardiovasc Surg 2018;24:173-179.

5. Litke-Wager C., Delaney H., Mu T., Sawyer T., Impact of Task-Oriented Role Assignment on Neonatal Resuscitation Performance: A Simulation-Based Randomized Controlled Trail, Am J Perinatol 2020. doi: 10.1055/s-0039-3402751
6. Buljac-Samardzic M., Doekhie KD., van Wijngaarden JDH., Interventions to improve team effectiveness within health care: a systematic review of the past decade, Hum Resour Health 2020;18:2.