Case Reports Intensivistendagen 2014

1.

Disastrous consequences: Influenza A and pneumococcal co-infection

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Introduction: Pneumococcal infection after influenza A infection lead more frequently to an invasive pneumonia. We describe a patient with influenza A and a pneumococcal co-infection who developed a severe septic shock with cavitary pneumonia and multi organ failure. Early recognition and treatment of a co-infection is important in the prevention of a more serious course of the disease.

Case history: A 64-year-old female, was admitted to the ICU with respiratory failure en septic shock with multiple organ failure. Her medical history compromised atrial fibrillation and hyperthyroidism. She suffered from fever, coughing and dyspnoea which started one week before admission. The chest radiograph showed bilateral infiltrates (*figure 1*).

Diagnostic tests showed positive PCR for pneumococcus and influenza A. Complementary blood cultures showed *Streptococcus pneumoniae.* Immediately after taking blood cultures and viral PCR, we started antibiotic and antiviral therapy.

Intubation, inotropic support and continuous venous hemofiltration were required.

The patient underwent prolonged postural drainage, prone position and several bronchoscopies treating sputum retention. A computed tomography of the chest revealed cavitating pneumonia (*figure 2*).

After a few weeks the patient recovered from the multiple organ failure and started weaning from the ventilator. She was discharged from the ICU after two months of admission (*figure 3*).

After two months of rehabilitation the patient restarted her work as a nurse.

Discussion: Approximately 250.000-500.000 people die of influenza worldwide, of which a significant fraction of influenza deaths can be attributed to bacterial infections, either during, or closely following influenza infection. An important influenza co-infection is pneumonia, caused primarily by *Streptococcus pneumonia*.[1]

Although the mechanisms underlying the association between the severity of influenza and pneumococcal infections are still poorly understood, there are a number of mechanisms involved. Viral infections facilitate bacterial colonization, adhesion and translocation through the epithelial barrier. In fact, clearing the way for bacterial disease. Pneumococcal infection after influenza infection lead more frequently to invasive pneumonia. In conclusion, it is clinically relevant to start with the combination of antiviral and antibacterial treatment in case of suspicion of a co-infection. This is of particular importance during the influenza season.[2] The co-treatment is only beneficial when the antiviral therapy is started within the first few days of onset of pneumonia. This can prevent the occurrence of a serious invasive pneumonia.[1]

Conclusion: This case showed a severe cavitary pneumonia following Influenza A and pneumococcal co-infection. We successfully treated this patient with prompt administration of antibiotics combined with antiviral therapy. Nevertheless a cavitary pneumonia developed with a prolonged treatment on the ICU. In conclusion, doctors should be aware of the fulminant course of the disease due to influenza and pneumococcal co-infection.

Figure 1. Chest radiograph at admission showed bilateral effusion

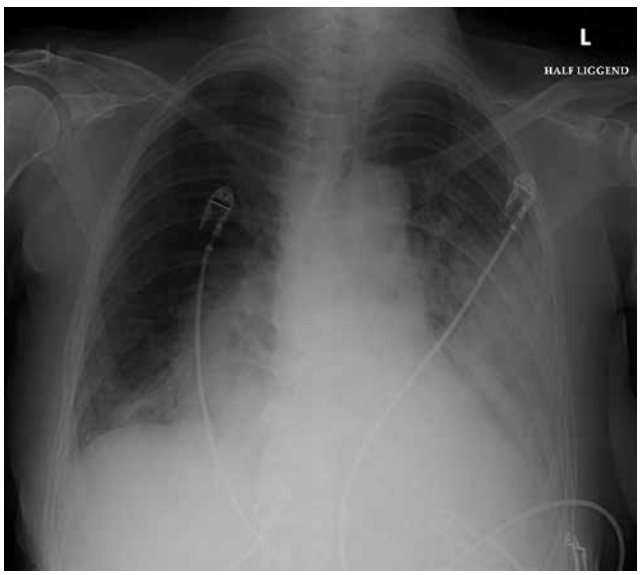


Figure 2. Computed Tomography: severe cavitary lung destruction air-fluid levels: confirmed lung abscesses

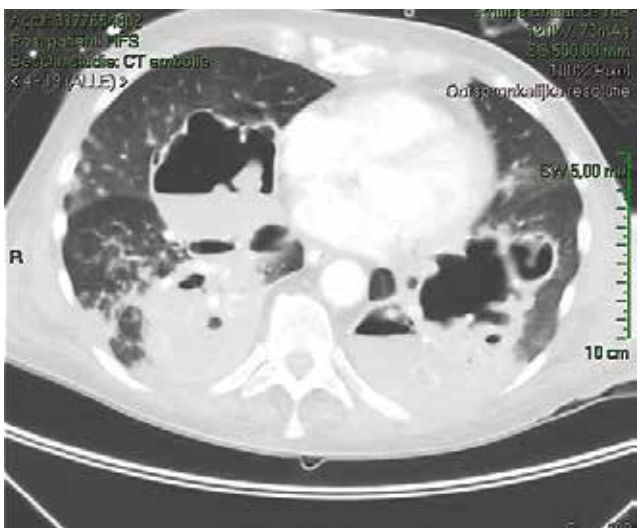
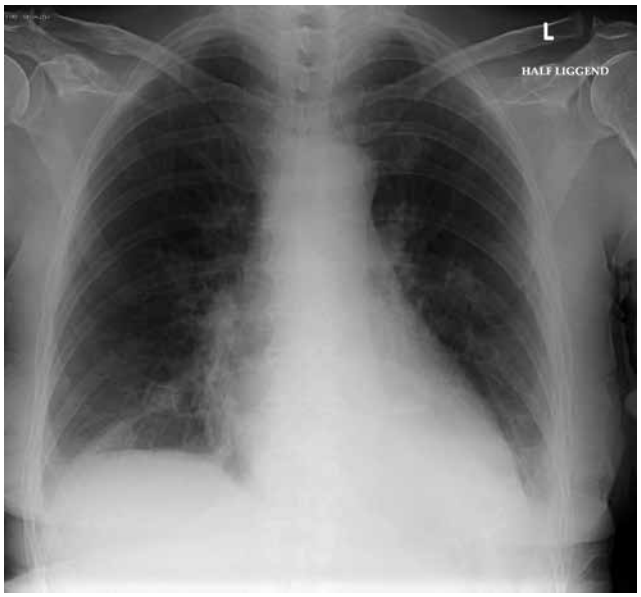


Figure 3. Chest radiograph at discharge: improved with only a few abnormalities left



References

1. Shresta S, Foxman B, Dawid S, et al. Time and dose-dependent risk of pneumococcal pneumonia following influenza: a model for within-host interaction between influenza and *Streptococcus pneumoniae*. J R Soc Interface. 10:20130233.
2. Hament J, Kimpen JLL, Fleer A, Wolfs TFW. Respiratory viral infection predisposing for bacterial disease: a concise review. FEMS Immunology and Medical Microbiology 26 (1999) 189-95.

2.

Hypokalemic paralysis and profound metabolic acidosis in a woman with Sjögren's disease

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Case: A 52-year-old woman presented with a rapid progressive proximal muscle weakness in both upper (Medical Research Council (MRC) grade 3-4) and lower (MRC 2) extremities and neck flexors (MRC 3) as well, since last week. She also documented anorexia, weight loss and constipation, for which she had used laxatives last week but abuse was not suspected. She was diagnosed with Sjögren's Syndrome (SS) four months before. The diagnosis was based on xerostomia, keratoconjunctivitis sicca, a positive Schirmer test and positive antinuclear antibodies with both positive anti-Ro/SSA and anti-La/SSB.

The laboratory results and the ECG on admission are shown in *table 1* and *figure 1*. Remarkable was the severe hypokalemia (1.5 mmol/l), the profound non-aniongap acidosis (bicarbonate 4.6 mmol/l, anion gap 13.6 mmol/L, albumin 41 g/l) and elevated creatine kinase (CK). Despite the profound metabolic acidosis, urine pH was 6.5. The anion gap in urine was 31 mmol/L with a urine osmolgap of 14 mOsm/kg.

Both the urinary anion gap and the urine osmolgap suggested the inability to produce ammonium. In combination with low bicarbonate and hypokalaemia, renal tubular acidosis (RTA) type I was diagnosed.

The woman received high doses intravenous potassium via a central venous catheter and when serum level was raised above 3.0 mmol/L, supplementation of bicarbonate was added. Her myopathy recovered soon after normalization of serum potassium and subsequently she was successfully treated with potassium citrate tablets (tid 1500 mg).

Discussion: Distal (type 1) RTA is an uncommon disorder, particularly in adults. The primary defect is impaired distal acidification with reduced urinary ammonium excretion. The hallmark features of RTA are hyperchloremic metabolic acidosis with a normal anion gap and a urine pH above 5.5, with or without associated defects in potassium homeostasis. The major causes of distal RTA in adults are autoimmune diseases, e.g. SS (*table 2*).[1] In up to 25 per cent of patients with SS, interstitial nephritis with a defect in distal acidification occurs.[2] The underlying mechanism is incompletely understood.

Differential diagnosis included multiple myeloma, but in our patient a recent protein electrophoresis was normal. This is important, because particularly haematological malignancies are more frequent in patients with SS and can also cause RTA type I.

Differential diagnosis of normal anion gap (hyperchloremic) metabolic acidosis also included diarrhea, but there was no history of diarrhea or intoxication in this patient who did not use diuretics. Normal anion gap metabolic acidosis due to laxative abuse exists in the presence of normal distal acidification, and hence it was unlikely to be the case of acidosis in this case.

The aim of therapy is to achieve a relatively normal serum bicarbonate concentration. Adults with distal RTA can be treated with 1 to 2 mEq/kg/day sodium bicarbonate or sodium citrate. Potassium citrate is indicated when hypokalaemia persists despite correction of the serum bicarbonate, like in this case.

Conclusion: Although RTA is a rare disorder, it is quite common in SS patients and these patients can present with profound hypokalaemia and metabolic acidosis.

Figure 1. ECG, showing characteristic changes; depression of the ST segment and a prolonged QT interval

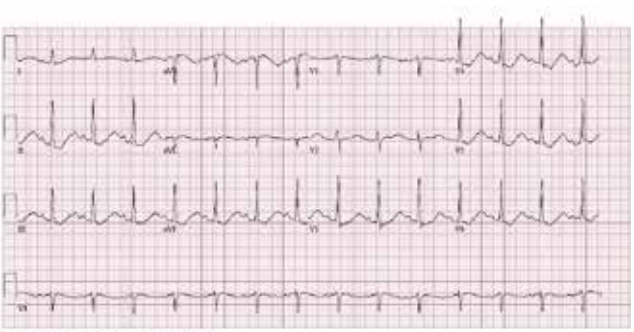

Table 1. Laboratory results

LABORATORY TEST	VALUE	REFERENCE VALUE
C-reactive protein (CRP)	17 mg/L	< 10 mg/L
ESR	50 mm/hr	2-12 mm/hr
Hemoglobin	6.6 mmol/L	7.0-9.2 mmol/L
MCV	87 fl	82-98 fl
Leucocytes count	13.4 x109/L	4.0-10.0 x109/L
Sodium	134 mmol/L	135-145 mmol/L
Potassium	**1.5 mmol/L**	3.5-5.0 mmol/L
Urea	11 mmol/L	2.5-6.4 mmol/L
Creatinin	173 µmol/L	44-80 µmol/L
Chloride	116 mmol/L	98-108 mmol/L
Calcium	2.25 mmol/L	2.10-2.55 mmol/L
Magnesium	1.12 mmol/L	0.65-1.05 mmol/L
Gamma-glutamyltransferase (GGT)	20 U/L	< 38 U/IL
Alkaline phosphatase (ALP)	50 U/L	< 98 U/L
Alanine aminotransferase (ALT)	40 U/L	< 34 U/L
Aspartate aminotransferase (AST)	80 U/L	< 31 U/L
Lactase dehydrogenase (LDH)	167 U/L	< 220 U/L
Creatine kinase	**1264 U/L**	< 145 U/L
Albumin	41 g/L	35-55 g/L
Glucose	5.9 mmol/L	3.0-7.0 mmol/L
TSH	3.37 mU/L	0.35-4.50 mU/L
pH	7.22	7.35-7.45
pCO2	1.5 kPa	4.7-6.4 kPa
pO2	16.0 kPa	10.0-13.3 kPa
Bicarbonate	**4.6 mmol/L**	22-29 mmol/L
Base excess	-20.6 mmol/L	-3.0-3.0 mmol/L
Urinary pH	6.5	4.0-8.0
Urinary sodium	14 mmol/L	25-135 mmol/L
Urinary potassium	12 mmol/L	16-67 mmol/L
Urinary chloride	13 mmol/L	110-250 mmol/L
glucose in urine	negative	

Table 2. Major causes of distal renal tubular acidosis (type 1)

Primary	
Idiopathic (sporadic)	
Familial	
Autosomal dominant	mainly due to mutations causing defects in the kidney anion exchanger – kAE1 in distal tubule intercalated cells.
Autosomal recessive	Mainly due to mutations causing defects in V-ATPase in distal tubule intercalated cells.
Secondary	
Autoimmune disorders • Sjogren's syndrome • Autoimmune hepatitis/primary biliary cirrhosis • Systemic lupus erythematosus • Rheumatoid arthritis	
Drugs • Lithium • Amphotericin B • Toluene inhalation • Ifosfamide	
Hypercalciuric conditions • Sarcoidosis • Hyperparathyroidism • Vitamin D intoxication • Idiopathic hypercalciuria	
Other • Sickle cell disease • Obstructive uropathy • Renal transplant rejection • Wilson's disease • Medullary sponge kidney	

References

1. van Daele PL, Zanen AL, de Ronde W, et al. Severe hypokalaemia with paralysis in a patient with distal renal tubular acidosis as an initial expression of Sjögren's syndrome. [Ernstige hypokaliëmie met paralyse bij een patiënte met een distale renale tubulaire acidose als eerste uiting van het syndroom van Sjögren]. Ned Tijdschr Geneeskd. 2002;146;218-21.
2. Ren H, Wang WM, Chen XN, et al. Renal involvement and follow up of 130 patients with primary Sjogren's syndrome. J Rheumatol. 2008;35:278-84.

3.

The powerful double-edged sword of thrombolysis

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Background: We present a case in which the direct therapeutic effects of thrombolysis in massive pulmonary embolism are made visible, clinically as well as by diagnostic imaging and invasive haemodynamic measurements. Thereby, we show the feared side effect, severe bleeding, which unfortunately occurred.

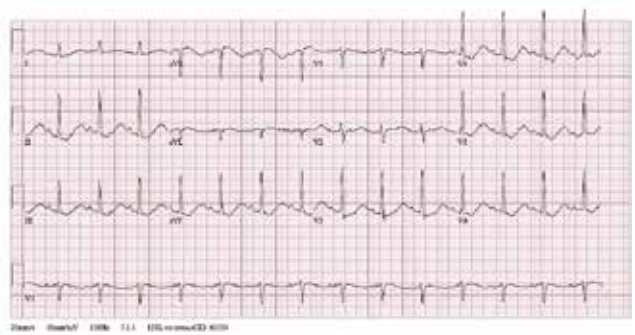
Case: An 82-year-old diabetic male was admitted to the ICU with a hyperosmolar hyperglycemic syndrome, luxated by prednisone use for a recent myasthenic crisis. He was treated with intravenous fluids, insulin and low molecular weight heparin (LMWH) in a prophylactic dose. Glucose and electrolytes normalized and no myasthenic crisis occurred. Patient was hypoxemic at admission, but further clinical examination and chest X-ray (CXR) were non-diagnostic. The next day he became severely hypoxic and haemodynamically unstable and therefore invasive mechanical ventilation was started. An electrocardiogram (ECG) showed a sinus tachycardia with S1Q3T3 pattern en incomplete RBBB (*figure 1*) and beds ide echocardiography revealed a dilated right ventricle. Pulmonary embolism (PE) was suspected and subsequently computed tomography (CT) pulmonary angiography confirmed extended saddle embolisms (*figure 2a*), with signs of severe right ventricular dilatation and left sided shift of the interventricular septum (RV/LV diameter ratio 2.8). The patient was treated with alteplase (rtPA), in the absence of any contraindications to thrombolysis, and subsequent LMWH in therapeutic dose. Significantly increased dead space ventilation with a Vd/Vt ratio of 70%,normalized after thrombolysis (about 24%).

The patient recovered well, but suffered an arterial bleeding after radial artery puncture. He was resuscitated with crystalloids, erythrocytes, platelets and Fresh Frozen Plasma. The LMWH was discontinued for the time coming. Nevertheless, he also developed a large hematoma in the right pectoral muscle (*figure 2b*). Since no extravasation of contrast was seen with CT imaging, no endovascular intervention was performed. Notably on CT, the large pulmonary embolism was almost completely dissolved and pulmonary artery catheterization measurements, after resuscitation, showed no elevated pulmonary artery pressures (*figure 3*). The next week, patient developed a ventilator-associated pneumonia (VAP) with refractory septic shock and eventually he died.

Discussion: Since it is generally known that the adverse effects of thrombolytic therapy can be devastating, the indication and potential benefits must be carefully weighed against the risks in each patient. Contraindications to thrombolytic therapy are recent (intracranial) surgery or head trauma, an active intracranial neoplasm, a history of stroke, severe hypertension, an active or recent internal bleeding, bleeding diathesis and thrombocytopenia.

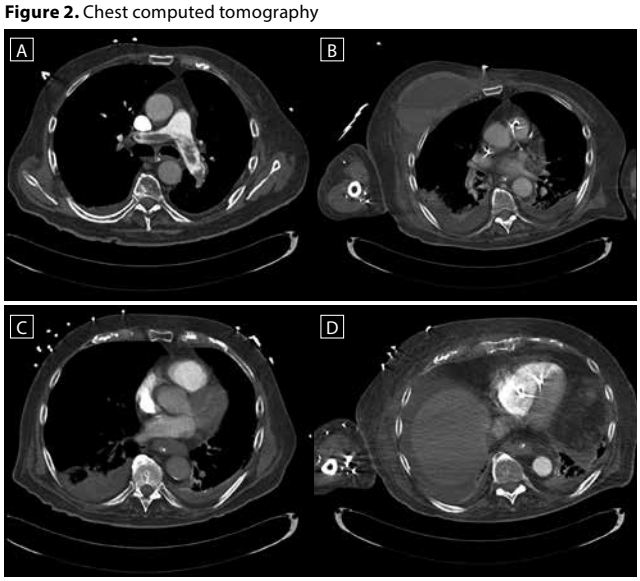
Although thrombolytic therapy can be lifesaving and leads to early haemodynamic improvement, it has not been proven to improve mortality in unselected patients.[1] However, in patients with major pulmonary embolism who are haemodynamically compromised, a consistent trend toward improved mortality has been repeatedly found. But as said, with a significant increased risk for major bleeding, compared to anticoagulation alone.[2]

Figure 1. ECG



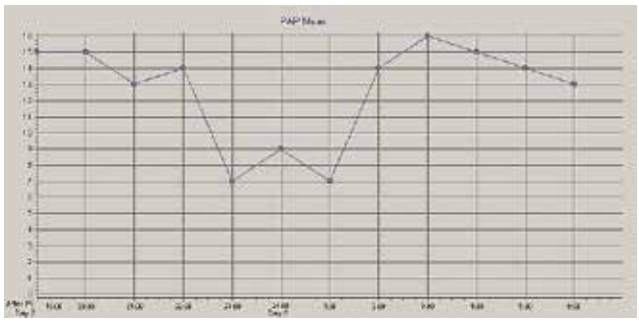
Typical ECG withS1Q3T3 pattern en incomplete RBBB

Figure 2. Chest computed tomography



Computed tomography scan of the chest, before (a. and c.) and after (b. and d.) thrombolysis.
Figure a & c: show the saddle embolus with concomitant right ventricular (RV) dilatation en shift of the septum to the left. The calculated RV/LV diameter ratio is significantly elevated; 2.8.
Figure b & d: show that the saddle embolus is almost completely dissolved after thrombolysis and the calculated RV/LV ratio decreased (although it was still elevated; 1.16). Thereby, it shows a large hematoma of 14 x 8 cm underneath the right M. Pectoralis.

Figure 3. Invasive haemodynamic measurements



Graphic with mean pulmonary artery pressures (PAP) measured by Swan-Ganz catheter one week after thrombolysis.

Conclusion: This case demonstrates the effectiveness of thrombolytic therapy in predominantly dissolving massive pulmonary embolism with normalization of PAP pressures, but at cost of the feared side effect. Because of this massive bleeding risk and the absence of strong evidence of mortality improvement, thrombolytic therapy should be used only in carefully selected cases.

References

1. Wan S, Quinlan DJ, Agnelli G, Eikelboom JW. Thrombolysis compared with heparin for the initial treatment of pulmonary embolism: a meta-analysis of the randomized controlled trials. Circulation. 2004;110:744.
2. Kearon C, Akl EA, Comerota AJ, et al. American College of Chest Physicians. Antithrombotic therapy for VTE disease: Antithrombotic Therapy and Prevention of Thrombosis, 9th ed: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines. Chest 2012;141:e419S.

4.

Giant-cell myocarditis – a giant challenge

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Introduction: Giant-cell myocarditis (GCM) is a rare disease carrying an unfavourable prognosis. A majority develops progressive heart failure and life-threatening arrhythmias. Thus, a timely diagnosis is essential and can only be obtained by myocardial biopsy. If medical therapy fails, only cardiac mechanical support and transplantation can be considered, although their therapeutic benefit in GCM remains to be debated. Here, we describe the clinical challenges one faces when managing a patient with GCM.

Case: A 22-year-old male without any medical history was referred in cardiogenic shock after complaining of general malaise for days. After stabilization upon medical treatment and routine diagnostics, right-ventricular endomyocardial biopsies revealed GCM. Yet, despite immunosuppressive therapy (steroids, cyclosporine and azathioprine) and intensified heart failure therapy, cardiac contractility deteriorated, necessitating the implantation of a bi-ventricular assist device (Thoratec®CentriMag®). Weaning from mechanical support was impossible, and 6 weeks after presentation cardiac transplantation was performed, allowing clinical discharge (day 65). Follow-up

biopsies revealed GCM recurrence 2 months after transplantation in the absence of heart failure and was successfully treated with methylprednisolon (3 days) and intensified prednisolone therapy.

Discussion: This exceptional case of recent-onset, unexplained heart failure emphasizes the incremental diagnostic value of an early endomyocardial biopsy (EMB). The diagnosis of GCM cannot be established by any other diagnostic means. Therefore, it is of utmost importance to consider EMB early in the diagnostic work-up.

Although GCM is a rare, idiopathic disease, the diagnosis has important therapeutic implications, i.e., the initiation of immunosuppressants. In our case, combined immunosuppression and adjuvant medical unloading did not reverse clinical deterioration due to terminal bi-ventricular failure. Generally, cardiac transplantation is the treatment of choice in refractory GCM, but this therapeutic option is limited by the paucity of donor organs. Alternatively, short-term mechanical support as a 'bridge-to-transplantation' can be considered, as we did. Yet, bi-ventricular support is cumbersome and bears significant risks of bleeding, thromboembolism and infection. Moreover, modern bi-ventricular short-term support devices, e.g., veno-arterial extracorporeal membrane oxygenation orextracorporeal centrifugal pumps (Thoratec® CentriMag®), are designed for limited use (a few weeks). Thus, the time interval to transplantation may exceed the durability of the short-term device. In contrast, long-termpulsatile bi-ventricular support devices disqualify for a 'high-urgency' waiting list and carry unacceptably high complication rates including irreversible pulmonary hypertension after months of support which precludes cardiac transplantation.

Moreover, GCM is well-known to potentially recur in the transplanted heart, but has been reported to allow successful treatment by adapting immunosuppressants, as in this case.

Conclusion: Severe, idiopathic heart failure of recent onset should prompt EMB in order to establish a diagnosis. Rarely, GCM may be encountered which carries a poor prognosis but may be treated by employing a challenging therapeutic strategy combining medical and mechanical cardiac support towards transplantation.

References

1. Cooper LT Jr, et al. Idiopathic Giant-Cell Myocarditis – Natural History and Treatment. N Engl J Med. 1997;336:1860-6.
2. Murray LK, et al. Ventricular assist device support as a bridge to heart transplantation in patients with giant cell myocarditis. Eur J Heart Fail. 2012;14:312-8.

5.

Shock after colonoscopy

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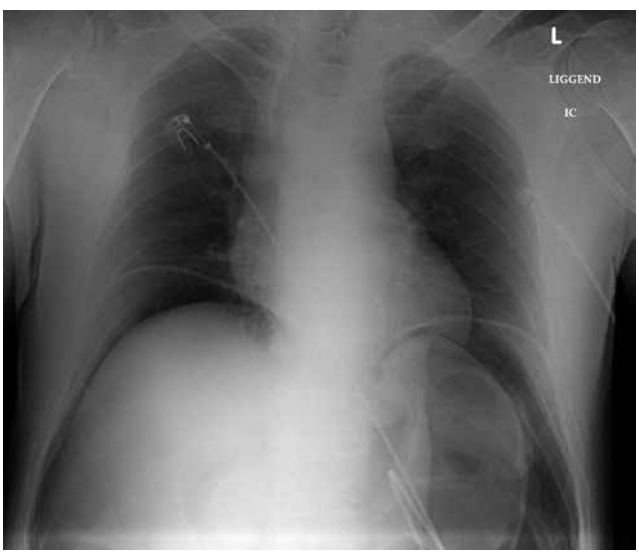
Introduction: An increase in intra-abdominal-pressure (IAP) can lead to an abdominal compartiment syndrome (ACS) with obstructive shock en multiple organ failure. One of the causes is a pneumoperitoneum. We present a case where a patient is referred to the ICU with obstructive shock following colonoscopy.

Case: A 69-year-old man was admitted to the ICU with tachypnea, hypotension and abdominal discomfort following gastro- and colonoscopy. He had a history of urolithiasis, DVT and non-insulin-dependent type 2 diabetes mellitus. He was referred to the hospital for analysis of microcytic anaemia. With gastroscopy no explanation was found, biopsies from duodenum where taken. During colonoscopy a narrowing colon tumor is observed, biopsies are taken and the tumor is marked proximal and distal of the tumor. Within a few hours the patients deteriorates with abdominal pain and shock and is admitted to the ICU. At physical examination we find extreme abdominal distension with redness of the lower limbs and an erection of the penis. Perforation or bleeding with obstructive shock is suspected. A radiograph of the chest shows a massive amount of intra-abdominal air (*figure 1*). With prompt needle decompression of the abdomen the patients circulation dramatically improved. Subsequent laparotomy show s a pneumoperitoneum with a perforation of the tumor. After sigmoid resection and antibiotics the patient recovered.

Figure 1. Plain radiograph of the chest: a massive amount of intrabdominal free air is striking



Discussion: ACS is defined as an increase in IAP which is associated with new organ failure or dysfunction.[1] Especially in ICU patients ACS is a known complication in trauma-patients, burn-victims, intra-abdominal surgery, polytransfusion, intra-abdominal bleeding and ascites and edema due infection, inflammation or malignancies.[1] We present a rare cause of ACS caused by perforation of a colontumor after biopsy during colonoscopy. Perforation during colonoscopy is a rare complication itself with an estimated incidence of 0.03% to 2%.[2] During colonoscopy insufflation of air is used to visualize the intestinal wall. If perforation occurs but not recognized, it can lead to a tension-pneumoperitoneum.[2] Probably the omentum acts as a one-way-valve preventing air to flow back into the intestine.

The use of needle-decompression is usefull as a salvage procedure

during resuscitation, although it has been described as sole therapy in one case.[2] This is well reflected by our case in which circulation dramatically improved after decompression by needle, before further surgical intervention was planned. In general colonic perforation is associated with high morbidity, up to 50%. Therefore surgical management is widely recommended.[2]

Conclusion: ACS as a result of perforation following colonoscopy is a rare but most serious complication. It should be considered in the patient in shock after colonoscopy. Endoscopists but intensivists as well, should be aware of this complication and its clinical signs. Although not a definitive therapy needle-decompression is quick, readily available and particular helpful in resuscitation of acute ACS due to pneumoperitoneum. Afterwards swift surgical intervention is still warranted.

References

3. Papavramidis TS, Marinis AD, Pliakos I, et al. Abdominal compartment syndrome-Intra abdominal hypertension: Defining, diagnosing, and managing. J Emerg Trauma Shock. 2011;4:279-91.
4. Souadka A, Mohsine R, Ifrine L, et al. Acute abdominal compartment syndrome complicating a colonoscopic perforation: a case report. J Med Case Reports. 2012;6:51.

6.

Unexplained hypoxia and Budd Chiari syndrome in a patient with antiphospholipid syndrome

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Case report: A 23-year-old woman was admitted for analysis and treatment of a suspected Budd-Chiari syndrome. Her medical history reported antiphospholipid syndrome, complicated by pulmonary emboli and thrombosis of the superior vena cava (SVC) and brachiocephalic vein in the previous year. The Budd-Chiari syndrome was caused by a suprahepatic inferior vena cava (IVC) thrombus. On admission she already complained of dyspnoea d'effort. A day after admission, there was progressive respiratory failure. She was transferred to the Intensive Care, where she soon needed to be intubated and ventilated. On suspicion of pneumonia, antibiotics were started. However, her condition did not improve and marked hypoxia persisted despite high FiO2 and ventilation pressures. A CT scan, with iodine contrast injected through the femoral vein, ruled out new pulmonary embolism, and showed only marginal pleural effusion and atelectasis, and no signs of interstitial lung disease.

The differential diagnosis, in the setting of Budd-Chiari syndrome, included portopulmonary hypertension, or shunting due to hepatopulmonary syndrome. However, pulmonary artery catheterization revealed normal pressures. Contrast echocardiography was unremarkable and showed no signs of a right-left shunt. In addition, (99 m)Technetium-macro aggregated albumin (MAA) perfusion scanning showed 22% MAA-capture in cerebrum/kidney, possibly indicating pulmonary shunting.

Upon further respiratory deterioration over time, a CT scan was repeated to rule out new pulmonary embolism. This time the iodine contrast was administered through the right arm, revealing a totally occluded SVC and a severe stenosis of the distal azygos vein due to thrombosis. There were extensive chest wall collaterals, as well as collaterals that appeared to have a close relation with the right pulmonary veins. There was hardly any contrast in the right atrium and pulmonary arteries, but dense contrast enhancement in the right pulmonary veins, left atrium and left ventricle, suggesting direct shunting between systemic veins and pulmonary veins based on a SVC syndrome, rather than hepatopulmonary syndrome. Based on these findings a stent was placed to dilate the azygos-SVC junction. In the same procedure, a stent was placed over the calcified stenosis in the IVC. Oxygenation markedly improved directly after stent placement and the patient was weaned from the ventilator within 24 hours. She was discharged to from the ICU 4 days after the procedure. The ascites resolved completely within two weeks.

This case illustrates the different diagnostic steps in a patient with refractory hypoxia. The diagnosis was only made after (co-incidentally) altering the route of contrast administration. In the end, there was a relatively simple solution for the clinical problem.

7.

Massive airembolus after LVAD placement in a patient with dilating end-stage heart failure

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Background: In the treatment of end-stage heart failure left ventricular assist devices (LVAD) are well-known therapeutic options as a bridge to transplantation or even a bridge to destination in patients deteriorating under maximal inotropic therapy. Early complications include perioperative hemorrhage, air embolism, and right ventricular failure. Beyond the perioperative period, late complications consist primarily of infection, thromboembolism, and primary device failure.

In this case report we would like to share our experience in one of the severe complications that might appear after implantation of an LVAD.

Methods/Case: A 65-year-old female was admitted at our hospital suffering from dilating cardiomyopathy. In the previous decade she had been treated repeatedly for heart failure. Despite resynchronization therapy with pacemaker the estimated left ventricular function decreased from 25 to 10 %. Because of atrial fibrillation there was a progressive deterioration of right and left ventricular function, with pronounced clinical deterioration despite inotropic support. Patient underwent an aortic valve repair, a tricuspid valve repair because of severe aortic- and tricuspid

regurgitation and implantation of a Centrimag LVAD. After weaning from cardiopulmonary bypass, haemodynamic support was provided by low dose dobutamine, milrinone, amiodarone and high dose norepinephrine. At admittance to the Intensive Care Unit (ICU) patient showed signs of low cardiac output, which was corrected by fluid resuscitation. Trans esophageal ultrasound (TEE) showed no evidence of cardiac tamponade. Aproximally an hour after arrival at the ICU there was a sudden drop of cardiac output (CO). Measured blood flow dropped below 1 L/min, with preservation of LVAD rpm. Pulmonary artery catheter showed a drop of CO, with only a slight increase in CVD or PAP. Resuscitation started immediately by fluid therapy and increasing inotropic support. Patient was hypoxic with poor circulation of the extremities. Physical examination showed normal bilateral lung sounds. Inspection of the LVAD showed massive amounts of air in the cannula, with an airlock at the pump. Patient was immediately put in Trendelenburg position to try to avoid air-embolus to the brain.TEE showed marginal right- and left ventricular movement, with no evident opening of the aortic valve. There was a pronounced air-embolus in the ascending and descending aorta (*figure 1*). Re-thoracotomy performed on the ICU did not show a luxation of the apex-cannula. After advancing of the cannula into the left ventricle circulation swiftly restored. TEE showed increased right and left ventricular movement, with no evidence of outflow obstruction (*figure 2*). Negative pressure in the trabecular system of the left ventricle could have caused the aspiration of air in the apex-cannula. Patient recovered haemodynamically in the next days. She did not regain consciousness and developed epileptic seizures. CT-cerebrum showed widely spread ischemia. Patient died shortly after discontinuation of treatment.

Results/Conclusion: Air was aspirated into the LVAD. Causing airlock in the pump, outflow obstruction and fatal air-emboli to the brain. There was no evidence of displacement of the cannula during re-thoracotomy. Advancement of the apex-cannula caused swift haemodynamic stabilization. Special care should be taken positioning of the cannula and testing for air-emboli, because of possibly fatal consequences.

Reference

1. Igor D, et al. Management of Air Embolism during HeartMate® XVE Exchange. Tex Heart Inst J. 2007;34:19-22.

Figure 1. Air in the ascending aorta

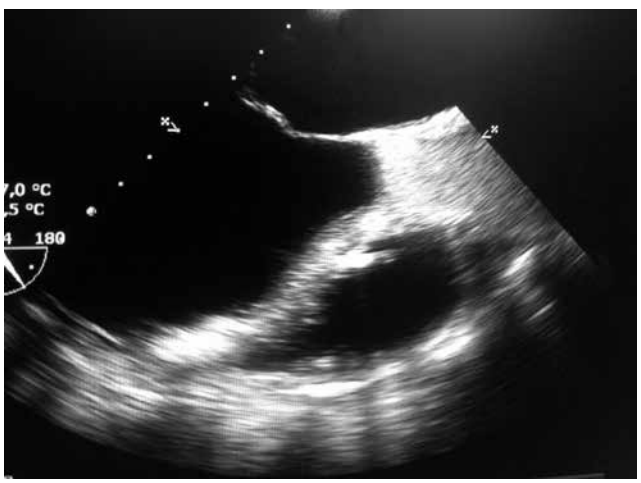
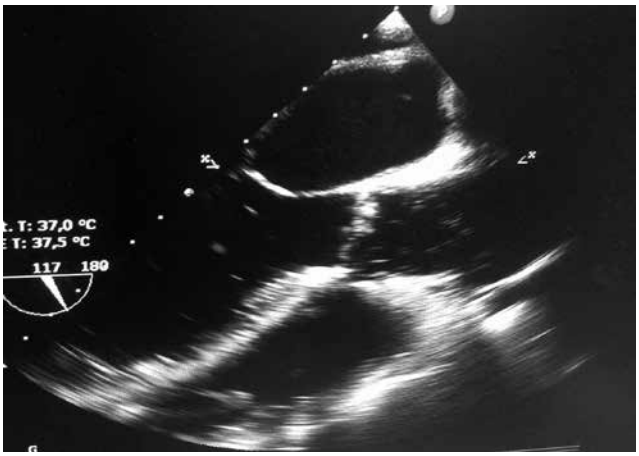


Figure 2. After repositioning of apex-cannula



8.

Blinded by septic shock

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Introduction: The majority of patients discharged from critical care experience problems with physical, non-physical and social functioning. We report on bilateral blindness as a rare and disabling complication in a young survivor of septic shock.

Case report: A 42-year-old male with a history of psoriatic arthritis, for which he used methotrexate, presented to our Emergency Department with abdominal discomfort two days after an inguinal hernia repair. Ultrasound showed abdominal wall hematoma and he was admitted to the surgical ward. Within 12 hours of admission he developed septic shock. The patient was admitted to the Intensive Care Unit (ICU), was resuscitated with fluids and vasopressors and treated with antibiotics (ceftriaxone and metronidazole). Emergency surgical exploration showed necrotizing fasciitis and aggressive surgical debridement was performed. Cultures revealed hemolytic Group A streptococcus and antibiotic treatment was switched to penicillin and clindamycin. Intravenous immunoglobulin was added for treatment of toxic shock syndrome.

The patient suffered from severe septic shock with multiple organ failure, including diffuse intravascular coagulation (DIC) and acute respiratory distress syndrome. High dose vasopressor therapy was needed (noradrenaline 2.7 mcg/kg/min and adrenaline 0.08 mcg/kg/min). After extubation he reported blindness with both eyes. His pupils were dilated and unresponsive to light. Ophthalmological evaluation revealed peripapillary retinal hemorrhages without signs of papillary edema. Magnetic resonance imaging showed fluid surrounding the optic nerves. Visually evoked potentials were negative. The patient was diagnosed with bilateral posterior ischemic optic neuropathy (PION). A course of steroids was considered to treat optic nerve edema but was found unsafe at that moment since he still had an active infection. Two weeks after diagnosing PION a course of dexamethasone was started

without improvement of vision. Follow-up ophthalmological examination showed regression of retinal hemorrhages but four months after diagnosis he still had no light perception or pupillary reaction to light and fundoscopy revealed bilateral optic nerve pallor.

Discussion: PION is a watershed infarction of the optic nerve and it is a rare but serious complication of critical illness. Blood supply to the posterior optic nerve is almost entirely dependent on the pial vasculature, which is very susceptible to ischemia. Factors associated with PION are hypotension, anemia, venous congestion, prone position, large blood loss, use of vasopressors and preexisting vaso-occlusive disease. Other causes of POIN are listed in *table 1*. A course of steroids can decrease optic nerve edema and thereby improve outcome. Spontaneous recovery or improvement of PION is unlikely.

The exact cause of PION in this case remains unclear. The patient suffered from severe septic shock with multiple organ failure and received high dose vasopressor therapy. However, he did not exhibit other signs of systemic hypoperfusion such as renal failure or gastrointestinal problems. He was never ventilated in prone position, did not suffer from excessive blood loss or anemia and was not known to have vaso-occlusive disease. The presence of retinal bleeds in the initial ophthalmologic examination leads us to think DIC played a contributing role in causing PION in our patient.

Conclusion: We report on a rare complication of septic shock: bilateral irreversible complete vision loss due to PION.

Reference

1. Lee LA, Nathens AB, Sires BS, et al. Blindness in the intensive care unit: possible role for vasopressors? Anesth Analg. 2005;100:192-5.

Table 1.

Etiology of PION
1) a combination of systemic hypotension and anemia (due to blood loss)
2) giant cell arteriitis, vasculitis (herpes zoster, polyarteritis nodosa, lupus erythematosis)
3) infiltrative causes
4) compression
5) idiopathic

9.

Ictal asystole: a rare complication of epilepsia

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Background: Epilepsy can be associated with a variety of cardiac arrhythmias. Ictal asystole (IA) is a rare, and potentially fatal complication.

Methods: A 32-year-old, 17 weeks pregnant patient was brought to the emergency room after return of spontaneous circulation after cardiac arrest. She had a history of partial complex epilepsy. She used carbamazepine which was stopped at the beginning of her pregnancy. After a seizure she remained unconscious and cardiopulmonary resuscitation (CPR) was started. The ambulance arrived after 10 minutes and continued CPR with a monitored asystole. She had return of spontaneous circulation and on arrival at the ER her blood pressure (BP) was 170/100 mmHg, pulse of 131/min. Her Glasgow Coma Scale (GCS) was 3 with normal brain stem reflexes. CT cerebrum showed diffuse swelling with compressed ventricles and a hypodensity of the right parietal lobe, considered to be preexistent. There were no signs of intracranial hemorrhage nor signs of brain stem compression (picture 1). Her blood work showed a bloodgas with a pH of 6,75 a pCO2 of 71 mmHg and a lactate of 16,6mmol/l. A transthoracic echocardiography (TTE) showed a hyperdynamic left ventricle without regional wall motion abnormalities and no valve abnormalities. There were no signs of pulmonary embolism on a CT angiogram. She was admitted to the ICU for therapeutic hypothermia according to protocol. After rewarming a somatosensory evoked potential (SSEP) was performed which was not conclusive due to technical difficulties so an electroencephalogram (EEG) was performed which showed burst suppression without reactivity. One day later the EEG showed an isoelectric trace, without any cortical activity and no signs of non-convulsive state. Treatment was discontinued and she died.

Results: Epileptic seizures are accompanied by cardiac arrhythmias 2% of which are IA or bradycardia. Schuele et al found an 0.27% incidence after evaluating 6825 cases of epilepsy.[1] Schuele used EEG and R-R interval monitoring and found asystole in 10 out of 6825 patients; eight in patients with temporal epilepsy and two of other origin.

Risk factors for IA are refractory epilepsy, most often temporal epilepsy, cardiac abnormalities and some anti-epileptic drugs(AED). There is some debate whether epilepsy triggers a respiratory arrest and concurrent hypoxia causing cerebral hypoperfusion and myocardial hypoxia and asystole or whether epilepsy alone is responsible through a autonomic response and discharge of the vagal centers in the medulla which leads to asystole resulting in cerebral hypoperfusion and atonia. This latter cardio-inhibitory pathway may be associated with temporal epilepsy.[1]

Treatment of IA or bradycardia requires better antiepileptic treatment and sometimes pacemaker implantation. Strzelczyk et al proposed the following algorithm.[2]

Conclusion: Ictal asystole is a rare complication of epilepsy with difficult etiology but is potentially fatal. In our patient the asystole was preceded by a seizure which probably led to cerebral hypoperfusion and ischemia of both the heart and brain. Surviving patients should receive aggressive treatment with AED, epilepsy surgery, pacemaker implantation or a combination of treatments.

References

1. Schuele SU, Bermeo AC, Alexopoulos AV, et al. Video-electrographic and clinical features in patients with ictal asystole. Neurology 2007;69:434-41.
2. Strzelczyk A, Cenusa M, Bauer S, et al. Management and long-term outcome in patients presenting with ictal asystole or bradycardia. Epilepsia. 2011;52:1160-7.

Picture 1.

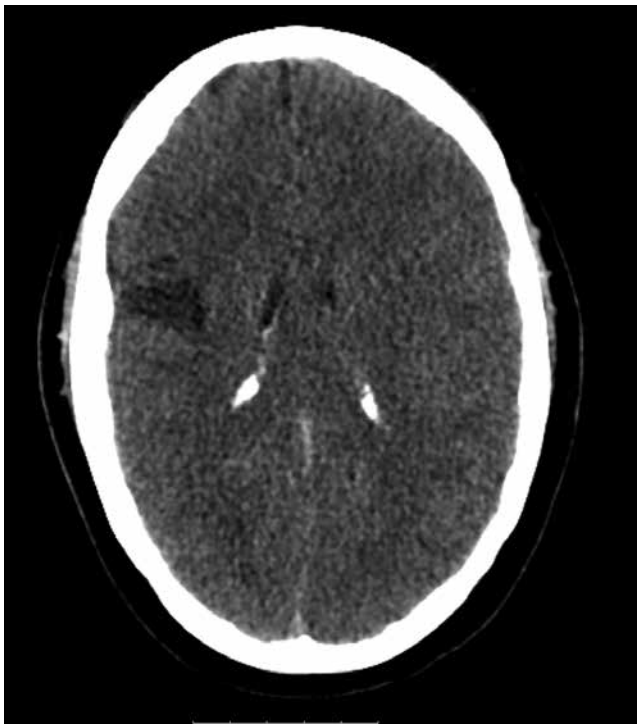

Figure 1.

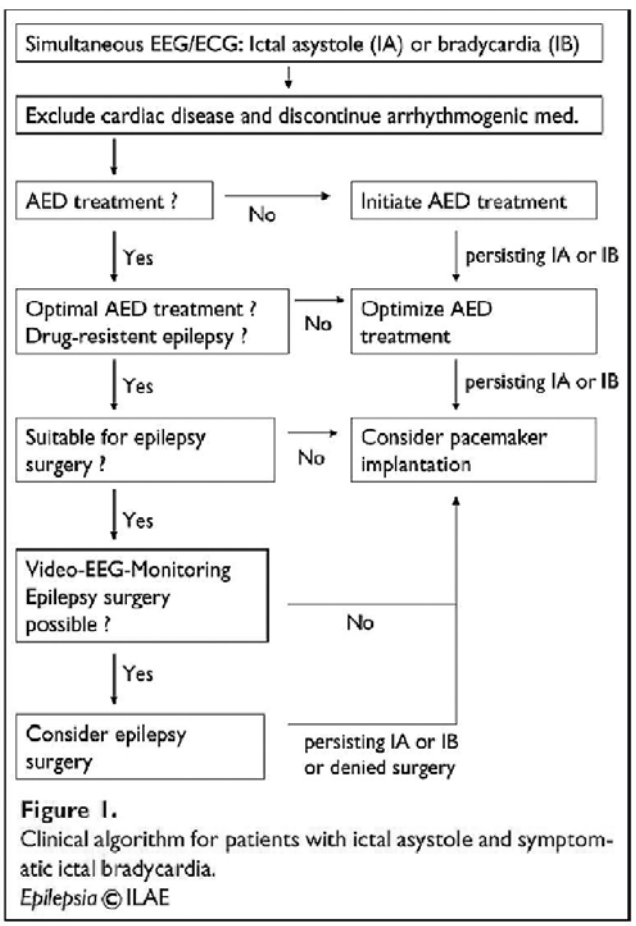

Figure 1.
Clinical algorithm for patients with ictal asystole and symptomatic ictal bradycardia.
Epilepsia © ILAE

10.

A case of fatal asthma after use of a dietary supplement containing creatine

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A 49-year-old man, with a history of mild allergic asthma necessitating occasional salbutamol inhalation therapy, developed a severe asthma attack some 30 minutes after intensive workout and a first-time ingestion of a protein shake containing creatine. Following self-administered incremental doses of salbutamol he was found unresponsive. On arrival of the paramedics asystole and respiratory arrest were diagnosed. Extensive successful resuscitation was initiated and the patient was transported to our hospital. The ventilation during transport was reported to be extremely difficult. On presentation at the emergency room we saw a patient with a striking rash covering all body parts. Capnography could not be obtained; the ET tube appeared to be placed in the oesophagus. After endotracheal reintubation the patient developed again pulseless electric activity combined with unmeasurable high EtCO2, high ventilation pressures and expiratory wheezing, suggesting a severe bronchusobstruction with dynamic hyperinflation. Initial bloodgas analysis revealed a severe combined respiratory and metabolic acidosis: pH of 6.78 pO_2 282 kPa pCO_2 161 kPa HCO_3^- 23.8 mmol/l BE-11.3 SaO_2 0.99. Leucocyte count showed marked eosinophilia. Cardiac massage was combined with adrenaline iv and nebulisation, ketamine infusion, magnesium sulphate iv, as well as salbutamol, ipratropium and steroids. The patient was mechanically ventilated with low frequency and prolonged expiratory time allowing normalisation of intrathoracic pressures. After restoration of circulation, mild therapeutic hypothermia was induced. Unfortunately, on rewarming patient was found to have absent brain stem reflexes and brain death was declared according to national protocol. Autopsy showed severe pulmonary oedema with eosinophilic and neutrophilic infiltrates in the alveoli and parabronchial tissue, consistent with fatal allergic asthma (*figure 1*).

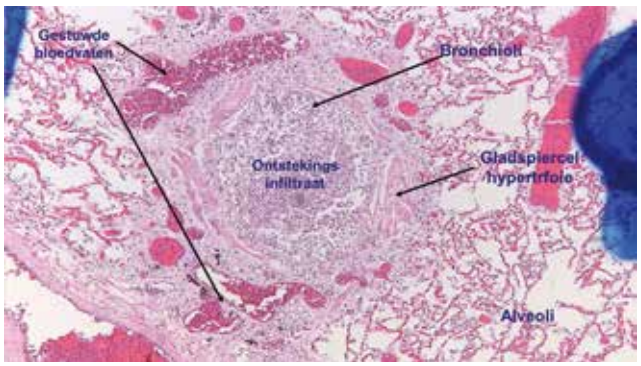
Although findings at autopsy could not be specifically linked to a distinctive allergenic cause, several clinical features in this case report are indicative for an allergic reaction after a first time ingestion of a creatine supplement. Creatine has been widely used as the most effective nutritional supplement with a proven ergogenic value in high-intensity exercise as well as longer duration exercise tasks, increasing muscle strength and lean body mass).[1] Also creatine supplementation is used in conditions where muscle wasting is a key feature like myopathies, neurodegenerative and brain disorders, heart failure and COPD.[1] Side effects seem to be limited to water retention; however when the maximal daily dose is exceeded, reports on kidney failure and hepatitis exist. In bodybuilder communities on the internet however, worsening of allergic asthmatic symptoms are frequently mentioned after use of creatine containing supplement; no case-report is yet available in the current literature on this matter. Vieira and co-workers [2] were on different occasions able to demonstrate in a model of sensitized mice that creatine supplementa-

tion exacerbates allergic inflammation, airway responsiveness, excessive mucus production and airway remodelling through activation of Th2 and IGF -1 pathways, all of which also have been proven to play a role in the development of human fatal asthma attacks. Therefore to our knowledge this is the first case in literature where the use of creatine supplementation can clinically be linked to a fatal allergic asthma.

References

1. Gualano B, Roschel H, Lancha AH, et al. In sickness and in health: the widespread application of creatine supplementation. Amino Acids. 2012;519-29.
2. Vieira RP, Duarte ACS, Claudino RC, et al. Creatine supplementation exacerbates allergic lung inflammation and airway remodeling in mice. Am J Respir Cell Mol Biol. 2007;37:660-7.

Figure 1. Cross-section of lungtissue, Clearly visible are congested vessels, inflammatory infiltrate and invasion of neutrophils and eosinophils



11.

Mirizzi syndrome mimicking malignancy: the right sequence in diagnostic interventions

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Introduction: The Mirizzi syndrome is a rare cause of obstructive jaundice and should be considered in the differential diagnosis of all patients with these signs. We describe a case of obstructive jaundice with severe complications due to diagnostic interventions. The sequence of these diagnostic and therapeutical interventions is important as complications could be prevented.

Case: A 55-year-old male patient with a history of Helicobacter Pylori gastritis presented with jaundice and itch since two weeks. On physical examination he was icteric without pain, the remainder was unremarkable. Blood test showed serum total bilirubine level of 147 umol/L, direct bilirubine 88 umol/L, alkaline phosphatase 241 U/L and γ-GT 97 U/L. Abdominal ultrasound revealed dilated intra- and extrahepatic biliary ducts, probably based on intraluminal obstruction of the common bile duct (CBD). The patient underwent an Endoscopic Retrograde Cholangiopancreatography (ERCP) which showed a proximal dilatation of the CBD (*figure 1*). Because of a stenosis in the medial part of the CBD an endoprothesis was placed.

A few hours after ERCP, the patient developed a hemorrhagic shock. He was admitted to the ICU where he received blood transfusion and fresh frozen plasma. Immediate CT angiography revealed a massive bleeding in the area of the papilla duodeni from one of the branches of the arteria hepatica propria (*figure 2*). Hemostatis was achieved by embolization. The next day our patient developed fever and further elevated bilirubine levels. Reassessment of the CT scan revealed a large, layered gallstone (size 3-4.5 cm) in the gallbladder neck as the cause of the obstruction (*figure 3*). The patient underwent laparoscopic cholecystectomy which was converted to an open procedure. A large gallstone was seen in the gallbladder neck that caused erosion of the lateral wall of the common bile duct (*figure 4*). Two days after surgery the patient was discharged from the ICU.

Discussion: Mirizzi's syndrome is characterized by a compression of the CBD or common hepatic duct caused by a gallstone which is impacted in the cystic duct or neck of the gallbladder. It causes obstruction and jaundice by an extrinsic compression of the CBD (type I) or may be accompanied by a cholecystobiliary fistula compromising the CBD wall (type II-IV). The primary radiologic tests to diagnose the Mirizzi syndrome are computed tomography combined with ERCP or MRCP.

Conclusion: In this case the patient went for ERCP after dilated biliary ducts were seen on abdominal sonography. If our patient went for computed tomography before ERCP a hemorrhagic shock could have been prevented. In conclusion, the right sequence of diagnostic and therapeutic interventions in obstructive jaundice should be accurately chosen since it may lead to severe complications.

References

1. Beltrán MA. Mirizzi syndrome: History, current knowledge and proposal of a simplified classification. World J Gastroenterol. 2012;18:4639-50.
2. Yun EJ, Choi CS, Yoon DY, et al. Combination of magnetic resonance cholangiopancreatography and computed tomography for preoperative diagnosis of the Mirizzi syndrome. J Comput Assist Tomogr. 2009;33:636-40.

Figure 1. ERCP with a proximal dilatation of the CBD and intrahepatic gallways with external compression in region of cystic duct

Figure 2. CT angiography reveals an active bleeding in the area of the papilla duodeni

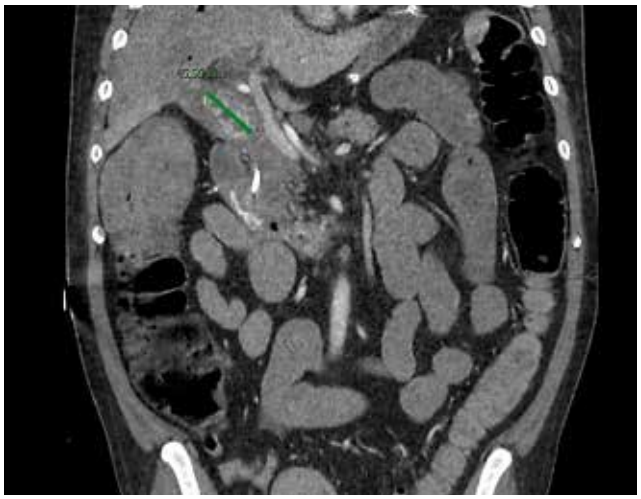

Figure 3. Computed Tomography showing a large, layered gall stone in the gallbladder neck, with compression of the common bile duct.

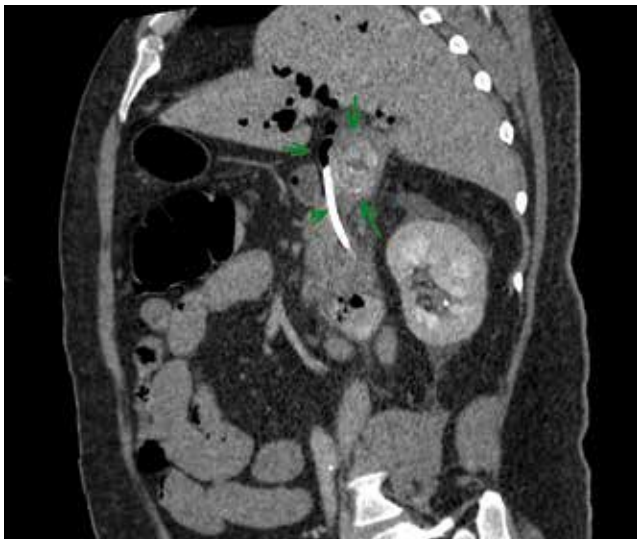

Figure 4. A large gallstone caused erosion of the lateral wall of the common bile duc

12.

Unexplained high lactate levels after long bone fractures: remember cerebral fat embolism syndrome

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Introduction: Cerebral fat embolism is an uncommon but serious complication of long-bone fracture. The classic fat embolism syndrome is characterized by the clinical triad of respiratory insufficiency, altered mental status and petechiae. Here we present two cases of cerebral fat embolism syndrome (CFES) and isolated high lactate levels without haemodynamic instability.

Patient A: A 26-year-old man was admitted to our hospital after being hit by a car. On arrival he had a maximal Glasgow Coma Score. Conventional radiology revealed fractures of both femura and a fracture of the left tibia. Treatment was entirely conservative. The patient was admitted to our intensive care unit (ICU) and within a few hour his consciousness decreased to an Eye Motor Verbal score of 3. Subsequently, his respiratory condition deteriorated quickly and the clinical picture and MRI (*figure 1*) were most suspect for CFES. Although there were no signs of sepsis, cardiac failure or ischemia, our patient had an persistent isolated lactate level between 4.9-8.2 mmol/L (*figure 2*). The patient died within 48 hours after admission to our hospital.

Patient B: A 19-year-old man presented with multiple trauma after a car accident. He had multiple fractures of the sacrum and femurs on both sides, a fracture of the right lower leg, and a tibia plateau fracture on the right. The patient underwent surgery and a total of three external fixtures were placed on both legs. The patient had a maximal Eye and Motor score while still on the ventilator. Several hours later he rapidly lost consciousness and within 20 minutes his Eye Motor Verbal score was E1M1Vtube. Almost at the same time the pulmonary condition of our patient deteriorated quickly. Given the neurological signs, in combination with the pulmonary deterioration, a diagnosis of fat embolism syndrome was suspected. Although there were no signs of sepsis, cardiac failure, or ischemia, our patient had an isolated lactate level of 3.1-5.5 mmol/L (*figure 1*). The patient died within 48 hours after admission to our hospital.

Discussion: Although the diagnosis CFES is difficult, several scoring systems have been developed to diagnose CFES. Here we suggest that lactate in the absence of haemodynamic instability could additionally be used in diagnosing CFES, adding it to its present criteria.

Although glucose is assumed to be the main energy source for all living tissue, there are indications that lactate, and not glucose, is preferentially metabolized by neurons in the brain. For example, lactate is an important metabolic precursor for cerebral gluconeogenesis and ATP under physiologic, but also pathological conditions.[1] At rest, the human brain releases a small amount of lactate. However, during cerebral activation, as seen during exercise, the plasma lactate increases and the brain takes up lactate in proportion to the arterial concentration.[2] It

is hypothesized that diffuse damage to the brain causes that neurons cannot use lactate whereas astrocytes and oligodendrocytes keep producing lactate with a net increase in lactate levels.

Possibly, adding lactate to the existing criteria for CFES, might increase the sensitivity of diagnosing CFES.

References

1. Stein NR, McArthur DL, Etchepare M, Vespa PM. Early cerebral metabolic crisis after TBI influences outcome despite adequate hemodynamic resuscitation. Neurocrit Care. 2012;17:49-57.
2. Quistorff B, Secher NH, Van Lieshout JJ. Lactate fuels the human brain during exercise. FASEB J. 2008;22:3443-9.

Figure 1. MRI

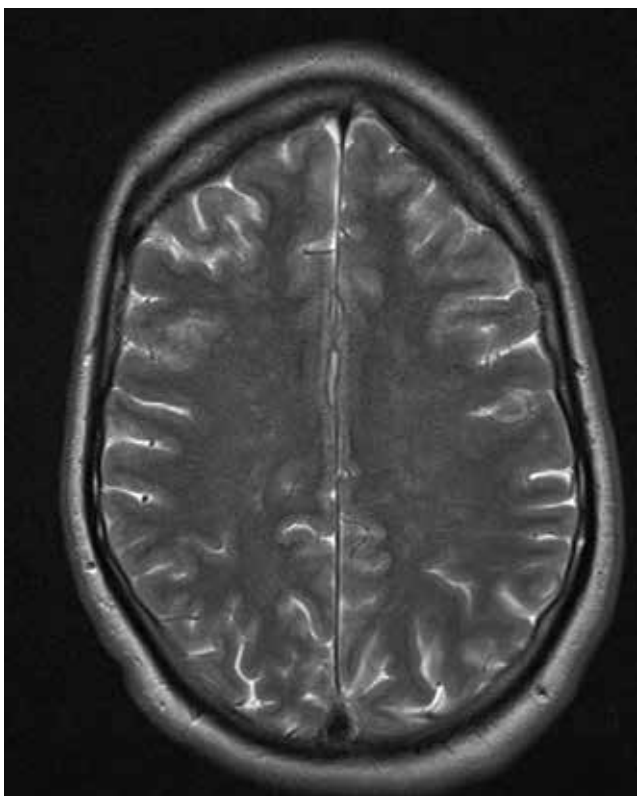
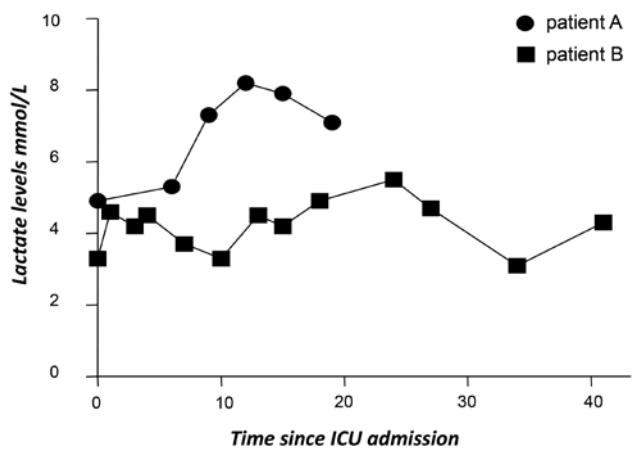


Figure 2. Lactate levels



13.

Diffuse alveolar hemorrhage, a rare but possible fatal complication after combined anticoagulant therapy for acute myocardial infarction

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Case report: Diffuse alveolar hemorrhage (DAH) is a possible life-threatening medical condition. Bland pulmonary hemorrhage due to elevated left ventricular end diastolic pressure is an underdiagnosed cause. Because of its low prevalence, DAH is a real diagnostic challenge. A 69-year-old man with a past medical history of myocardial infarction, presented to our emergency department (ED) after a witnessed out-of-hospital cardiac arrest while cycling. Basic life support was started immediately. Upon arrival of the ambulance, the presenting rhythm was ventricular fibrillation with subsequent return of spontaneous circulation after defibrillation. He remained comatose and arrived intubated in our ED. Electrocardiogram showed an atrial flutter without ST-elevation. Transthoracic echocardiography demonstrated an overall poor left ventricular function with an estimated ejection fraction of 30%. Prasugrel 60 mg, acetylsalicylic acid 300 mg and 5000 units of heparin were given because of the likelihood of a new ischemic event. A coronary angiogram revealed a culprit lesion in the right coronary artery. A bare metal stent was placed with good angiographic result. Because the patient vomited after administration of prasugrel in the ED, a loading dose of 600 mg clopidogrel was administered. After the PCI the patient was admitted to our intensive care unit. Therapeutic hypothermia was initiated. The respiratory situation deteriorated rapidly with a chest X-ray showing bilateral consolidations. Copious amounts of bloody secretions were aspirated from the endotracheal tube. Therapeutic hypothermia was stopped because of bradycardia and bleeding. Bronchoscopy showed diffuse bleeding in the distal regions of the bronchial tree suggesting DAH. Chest X-ray was repeated showing an increase in bilateral consolidations. The patient succumbed to refractory respiratory failure due to massive DAH.

In cardiac asthma there is a disruption of the layers of the alveolar-capillary unit due to elevated capillary hydrostatic pressures, a phenomenon also known as pulmonary capillary stress failure. A rise in capillary hydrostatic pressure favours the formation of edema in the interstitial compartment removed from the critical gas-exchanging regions. Once fluid forms in the interstitium, it is transported to the interlobular septae, the peribronchovascular space and finally to the hila and pleural space. Lymphatic vessels within these regions are highly recruitable and are able to increase the clearance of lung water by more than 10-fold. When this compensatory mechanism fails and all layers are disrupted, red blood cells may be seen traversing the alveolar-capillary membrane, resulting in the well-known pink frothy expectorations. Taking this pathophysiological finding into account, we believe that administration of anticoagulant therapy in patients with acute ischemic heart failure will increase the risk of diffuse alveolar hemorrhage, in this case a fatal complication.

This case suggests that administration of anticoagulant therapy after myocardial infarction can cause a fatal diffuse alveolar hemorrhage, especially if the patient is in acute ischemic heart failure. Diffuse alveolar hemorrhage can be mistaken for acute heart failure with similar radiological and clinical findings. Therapy should not only consist of treating acute heart failure, but cessation of implicated drugs and reversal of excess anticoagulation should be considered.

14.

Oxaliplatin induced multi organ failure: a condition intensivists should be aware of

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Background: Oxaliplatin is increasingly used in both adjuvant and palliative chemotherapy regimens for intestinal carcinoma. Consequently, more patients will be admitted to the intensive care unit (ICU) whilst on such a regimen. Common adverse effects include pancytopenia, mucositis, neuropathy and anaphylaxis. Less known are the potentially lethal hepatic effects, which will be illustrated in the following case.

Case: A 70-year-old female was transferred to our ICU from another hospital. Her medical history revealed non-metastasized rectum carcinoma, treated with neo-adjuvant chemotherapy and radical resection. Adjuvant treatment consisted of standard dose oxaliplatin and capecitabine. She was admitted with hypopotassaemia and metabolic acidosis due to mucositis. This was complicated by a pulseless electrical activity, resulting in an non-ST-elevation myocardial infarction. In the following days, sepsis of unknown origin with respiratory, renal and liver insufficiency with ascites developed. Laboratory results showed bilirubin 200 µmol/L, ASAT 160 U/L, ALAT 76 U/L, AF 779 IU/L and γ-GT 440 U/L. Moreover, thrombocytopenia with secondary intestinal bleeding occurred. She was intubated and platelet transfusion and antibiotics were administered. Gastroduodenoscopy showed multiple erosive ulcerations likely due to mucositis.

She was then transferred to our hospital because of lack of continuous venovenous haemodialysis capability in the referring center. Thrombopenia persisted at 23 10^9/L, in spite of transfusion. Diffuse intravasal coagulation was deemed unlikely with fibrinogen at 2.1g/L, PT 16,1s and aPTT 40s. Analysis of liver failure included extensive cultures, virus assays, computed tomography and ultrasound imaging to rule out metastasis or macrovascular obstruction and echocardiography to exclude both forward and backward failure. Besides the oxaliplatin, no other cause was found for this multiple organ failure. In spite of maximal supportive therapy, the patient died 6 days after admission. Unfortunately, postmortem confirmation of the diagnosis was declined by the family.

Discussion: Whereas the metabolic derangement due to diarrhea was probably caused by capecitabine related mucositis, the ensuing symptomatology is more likely due to oxaliplatin toxicity. Literature reports that oxaliplatin can cause sinusoidal injury syndrome (SOS), with subsequent, potentially fatal liver damage. Drug-induced thrombocytopenic purpura has been also described with oxaliplatin use. However, no fragmentocytes were found. The thrombocytopenia might also have been a drug-induced immune thrombocytopenia. This has however mainly been described whilst on oxaliplatin treatment, and recovers after discontinuation. Disseminated intravascular coagulation has also been shown after oxaliplatin use, but was excluded in this case. SOS therefore was the most likely diagnosis.

SOS has been described after oxaliplatin use. Most cases develop mild to moderate symptoms, and have a good prognosis. Severe SOS however, which accounts for 25-30% of cases, has a high mortality rate, especially if renal failure occurs. Treatment consists mainly of supportive care. If multi organ failure has not yet developed, anticoagulation may benefit outcome. Defibrotide, a polydeoxyribonucleotide, may benefit even those in severe SOS, but is not yet commonly available.

Conclusion: In case of prolonged thrombocytopenia and liver insufficiency after administration of oxalipatin, intensivists should be aware of the possibility of oxaliplatin toxicity as the causal entity.

References

1. Bautista, et al. Hypersensitivity reaction and acute immune-mediated thrombocytopenia from Oxaliplatin: two case reports and a review of the literature. Journal of Hematology and Oncology. 2010;3:12
2. Schouten van der Velden AP. Hepatic veno-occlusive disease after neo-adjuvant treatment of colorectal liver metastases with oxaliplatin: a lesson of the month. Eur J Surg Oncol. 2008;34:353-5.

15.

First Pieris Japonica intoxication described in a human

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Introduction: The Pieris Japonica (Japanese Pieris) plant is a member of the Ericaceae family, which also includes rhododendrons and azaleas. It is a shrub or small tree with oval to lanceolate leaves with finely serrated margins. They are more and more used as decoration in European gardening. They are also known to be toxic to animals, especially ruminants. These animals are often exposed to these plants in their grazing environment. A few reports have been published about intoxications of goats. As far as we are aware, this is the first report ever written about an intoxication in a human being.

Case: A 62-year-old woman was presented to the emergency room (ER) of the St Elisabeth Hospital with salivation, nausea, vomiting, bradycardia, hypotension and 'different behaviour'. Four ours before presentation, during her daily walk, she ate two branches of the Pieris Japonica plant. The woman is living in an environment for mentally disabled people. Her mentally performance is estimated at 0.5 years old.

In the ambulance her heart rate began slowing down to 40 beats per minute and her blood pressure dropped to 60/20. After one gift of atropine (0.3 mg) her blood pressure and heart rate stabilized. No further resuscitation was necessary.

On examination at the ER blood pressure was 124/86, heart rate was 110 R/A. Saturation was 98% (no extra oxygen), normal breath sounds. Her behavior was different to normal. Due to her pre-existent mentally performance a GCS was not possible. She didn't seem to be drowsy, both pupils were equal (no enlargement) and reactive to light. Further clinical evaluation showed no disturbances. Laboratory studies showed biochemical parameters within the normal range. The electrocardiogram revealed a sinus tachycardia of 107 beats/minute, a normal QRS axis, normal conduction times, no pathological Q waves and no ST segment changes.

Due to the amount of time between ingestion and presentation in the ER, no gastric lavage was performed. She was treated with activated coal and admitted to the Intensive Care Unit.

During her stay in the Intensive Care Unit she remained haemodynamically stable; no hypotension or arrhythmias were observed. The vomiting and salivation stopped. No neurological signs were observed and her behavior went back to normal.

The next day she was discharged to her own living environment.

Discussion: The poisonous principle of the plant is the acetylandromedol toxin. It binds to and modifies the sodium channels of cell membranes, leading to prolonged depolarization and excitation. It favors calcium movement into the cells and thereby results in a positive inotropic effect, similar to that of digitalis (although structurally unrelated). Bradycardia, hypotension and atrioventricular block are serious cardiovascular effects that may be lethal. As little as 0.2% of the body weight of leaves of the Pieris Japonica plant may be toxic and even lethal. We should be aware of the risks of this plant, particularly because of the low toxic dosage and the apparent increase in popularity in Europe.

16.

Colchicine poisoning; hesitate before extubate?

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Background: Colchicine is an anti-inflammatory drug primarily used for gout and familial Mediterranean fever. The drug has a low therapeutic index without a clear cut-off value between toxic and nontoxic dose. Acute colchicine poisoning is rare but associated with poor outcome. Colchicine is rapidly absorbed from the gastro intestinal tract and undergoes an extensive hepatic first-pass metabolism with significant enterohepatic re-circulation. Mean elimination half-life of oral colchicine is 4-16h but its half-life in leucocytes is reported to be 60 h. Colchicine binds to intracellular tubulin impairing the microtubular network. This predominantly affects cells with a high turnover rate and may subsequently lead to multi organ dysfunction revealing days after ingestion. The typical clinical course of colchicine poisoning is summarized in *table 1*. Therapy consists of gastrointestinal decontamination in the first phase and intensive supportive care.[1] Colchicine-specific Fab fragments are described to be effective, but are not commercially available.[2]

Case report: A 50-year-old woman with a history of gout and an unspecified psychiatric disorder was admitted comatose at an ED elsewhere 12-24h after an estimated ingestion of 15mg colchicine, 1600 mg clomipramine, 600 mg fenobarbital and three different benzodiazepines. She was intubated before transfer to our ICU. At admittance a E1M1V1 coma score persisted, without haemodynamically disturbances and unaffected oxygenation. Laboratory tests were unremarkable beside a creatine kinase (CK) of 3668 U/l. ECG showed normal sinus rhythm with prolonged QRS and QTc. Treatment was started with activated charcoal, magnesium sulphate and hyperhydration with saline and bicarbonate.

The next day patient fully awakened and appeared haemodynamic and respiratory normal with minimal support. CK level decreased and the conduction abnormalities normalized. Therefore it was decided to extubate.

Eight hours after extubation, 36-48h after ingestion, her condition suddenly deteriorated with shock, bilateral inspiratory crackels and severe hypoxemic respiratory failure not improving after applying non-invasive positive pressure ventilation. Chest X-ray showed bilateral alveolar consolidations compatible with (non-)cardiogenic lung edema. The patient was re-intubated and subsequently treated with lung protective ventilation in prone position. The following week she gradually improved although the ICU treatment was complicated by pneumonia, delirium and hypophosphatemia. On day 7 she was extubated and on day 9 she could be discharged from the hospital apparently without any residual symptoms.

Conclusion: Colchicine poisoning is a potential life-threatening condition. As our case report underlines, the initially mild clinical course may be misleading and acute deterioration may reveal. It is important to be aware of the subsequent clinical stages and patients should be closely monitored for several days after colchicine poisoning.

References

1. Finkelstein Y. Clin Toxicol; 2010;48:407.
2. Baud FJ. N Engl J Med. 1995;332:642.

Table 1. Clinical stages of colchicine poisoning.[1]

Stage	Time of onset	Features
Gastrointestinal phase	**0-24 hours** post-ingestion	• Nausea, vomiting, diarrhea, abdominal discomfort • Hypovolemia • Leukocytosis
Multi-organ failure phase	**1-7 days** post-ingestion	• **(Adult) Respiratory distress syndrome (ARDS)** • Cardiac arrhythmias, failure, arrest • Encephalopathy, brain edema, convulsions • Renal failure • Liver failure • Disseminated intravascular coagulation • Bone marrow suppression, pancytopenia • Metabolic acidosis, hypokalemia, hyponatremia, hypocalcemia, hypo(hyper)glycemia, hypophosphatemia • Myopathy, neuropathy • Secondary sepsis
Recovery phase	**7-21 days** post-ingestion	• Resolution of organ system derangements • Rebound leukocytosis • Alopecia

17.

Microscopic polyangiitis with primary central nervous system manifestations

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Background: Vasculitis presenting with central nervous system manifestations is a rare diagnosis in the ICU.

Methods (Case): A 41-year-old woman with a medical history of mild asthma developed sensory loss of her lower extremities after she was admitted to hospital with acute onset of pain in the interscapular area. Eight months earlier she experienced paraesthesias and sensory loss of her lower body, with no abnormalities detected by lumbar puncture or magnetic resonance imaging (MRI). Now, MRI of the spinal cord demonstrated a subdural hematoma at the thoracic level (Th4-7) (*figure 1*) and dexamethasone was started to treat the myelopathy. Two weeks later she developed paralysis of her right leg and progressive general weakness leading to respiratory insufficiency requiring mechanical ventilation. Consecutive MRI showed an additional hematoma at the cervical (C3-5) level as well as ischemic areas (corpus callosum, medulla oblongata) in the brain. One day later her state of consciousness decreased to a Glasgow Coma Scale of E4M4V1, with a fixed gaze to the right. A third MRI revealed multifocal microinfarcts with magnetic resonance angiography (MRA) demonstrating stenoses in multiple cerebral arteries (*figure 2*). Differential diagnosis included infective endocarditis, coagulation disorder, vasculitis and lymphoma. Diagnostic tests were performed accordingly. Broad-spectrum antibiotics were started after cultures had been taken. The next day she developed an exanthematous rash. Although uncertain, high dose methylprednisolone was started to treat the suspected vasculitis. In order to ascertain the diagnosis, biopsy of the affected brain tissue was done. In the following days her condition did not improve and renal function deteriorated. Haemodynamic instability occurred as a result of gastrointestinal bleeding. Unfortunately, none of the test results could confirm the presence of vasculitis until one week after ICU admission. P-ANCA was present and directed against myeloperoxidase (ANCA-MPO > 100IU/ml), indicative of microscopic polyangiitis (MPA). Treatment was optimised by pulse cyclophosphamide. Plasmapheresis and rituximab were added because of acute severe disease.

Results (discussion): MPA is a systemic vasculitis affecting small- and medium-sized blood vessels and is associated with myeloperoxidase-antineutrophil cytoplasm antibodies (MPO-ANCA).[1] Almost any organ system can be affected and therefore symptoms can vary. Although neurological involvement is not uncommon, this mainly concerns the peripheral nervous system.[1] The unusual presentation of MPA in our case led to delay in diagnosis and irresoluteness about the most correct therapy. Early recognition and prompt therapy of vasculitis is crucial to prevent serious morbidity and mortality. We would like to stress that CNS manifestations can be the first presentation of MPA. Because the majority of patients with MPA present with glomerulonephritis or lung haemorrhage,[1] clinical studies have focussed on this patient population and even here the role of plasma exchange or other adjunctive therapies remains unclear. Recently, rituximab was proven at least as effective as cyclophosphamide for the induction of remission in patients with severe disease.[2] Unfortunately, our patient only partially recovered from her cerebral ischaemic insults and remains severely disabled.

Conclusion: MPA is a systemic disorder that can present with primary central nervous system manifestations. Early recognition and therapy are critical.

Figure 1. A MRI of the spinal cord showing a hematoma at the thoracic level

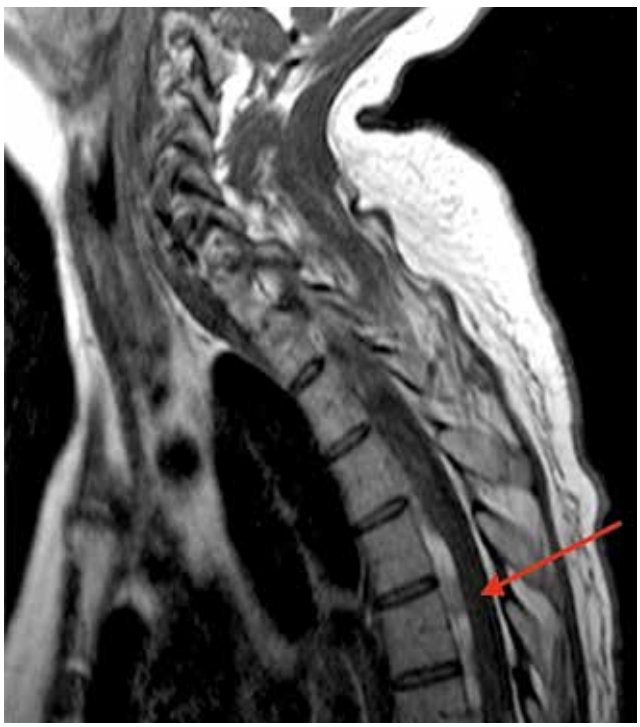
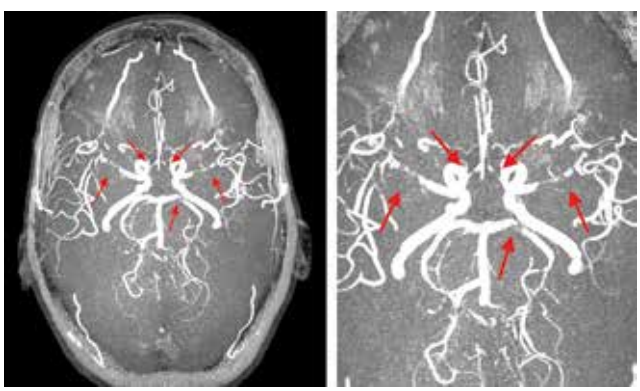


Figure 2. A MRA showing stenoses in multiple cerebral arteries



Thanks to doctor F.A.A. Mohamed Hoesein, Department of Radiology, University Medical Center Utrecht, Utrecht, The Netherlands

References

1. Chung SA, Seo P. Microscopic polyangiitis. Rheum Dis Clin North Am. 2010;36(3):545-58.
2. Specks U, et al. for the RAVE-ITN Research Group. Efficacy of remission-induction regimens for ANCA-associated vasculitis. N Engl J Med. 2013;369:417-27.

18.

Inadvertent placement of a Central Venous Catheter (CVC) in the subclavian artery. How to manage this problem?

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Background: Inadvertent puncture and catheterization of the subclavian artery is a known complication of infraclavicular catheterization for central venous access. Removal of the CVC without precautions can lead to severe complications.

Objectives: Management of inadvertent placement of a CVC in the subclavian artery.

Case report: A 74-year-old woman with an unremarkable medical history was recently diagnosed with an adenocarcinoma of the stomach. She received neo-adjuvantive chemotherapy and developed severe diarrhea. Patient was admitted to our ICU with deep hypovolemic shock due to dehydration. She was haemodynamically unstable and, besides aggressive rehydration, required inotropy. Insertion of a 7 French left subclavian CVC procedure was uncomplicated.

No excessive backflow was noticed. While the X-ray showed a 'correct' position of the CVC, an arterial pressure curve was registered. The consulted vascular surgeon advised not to remove the CVC and placement of a endovascular stent in a clinically more stable situation. It was not ruled out that the CVC had penetrated the vein prior entering the artery. Therefore, the use of a percutaneous closure device would not be sufficient. A 59 x 7 mm stent-graft was placed via the right femoral artery without compromising the vertebral and mammary artery. An indentation was dilated with a 7mm balloon. The CVC was removed without complications.

Discussion: Arterial catheterization of a subclavian CVC occurs in 1% of all infraclaviculair procedures. Catheterization in hypovolemic patients is difficult due to venous collapse. Risk factor for inadvertent arterial catheterization is, such as in our case, an emergency procedure.

Inadvertent arterial catheterization should be suspected when pulsatile backflow or local hematoma occurs. Control X-ray was performed and the position of the CVC was reported "correct" by the radiologist. If arterial catheterization is recognized, the catheter should be left in place and percutaneous, endovascular or surgical management should be considered. Removal of an arterial positioned CVC without precautions can lead to life-threatening complications. In our patient, stent placement was preferable because of its safety, low incidence of complications and good long-term results. A percutaneous closure device was contraindicated because of its high failure rate, complications and to avoid venous damage.

Ultrasound guidance reduces the risk of arterial puncture during placement of a CVC. However, the subclavian approach benefits less from ultrasound guidance. Therefore knowledge of correct management, when an accidental arterial catheterization occurs, is of great clinical importance.

Conclusion: Inadvertent arterial placement of a subclavian CVC is associated with emergency procedures. If arterial catheterization is recognized the catheter should be left in place. A systematic approach is necessary for planning the best treatment; endovascular, surgical or percutaneous.

References

1. Ruesch S, Walder B, Tramèr MR. Complications of central venous catheters: internal jugular versus subclavian access--a systematic review. Crit Care Med. 2002;30:454-60.
2. Polderman KH, Girbes AJ. Central venous catheter use. Part 1: mechanical complications. Intensive Care Med. 2002;28:1-17. Epub 2001 Dec 4.
3. Pikwer A. Acosta S., Kolbel T., Malina M., Sonessen B., Akeson J. Management of inadvertent arterial catheterization associated with central venous access procedures. Eur J Vasc Endovasc Surg 2009. 38, 707-714

Figure 1. False correct position of the CVC

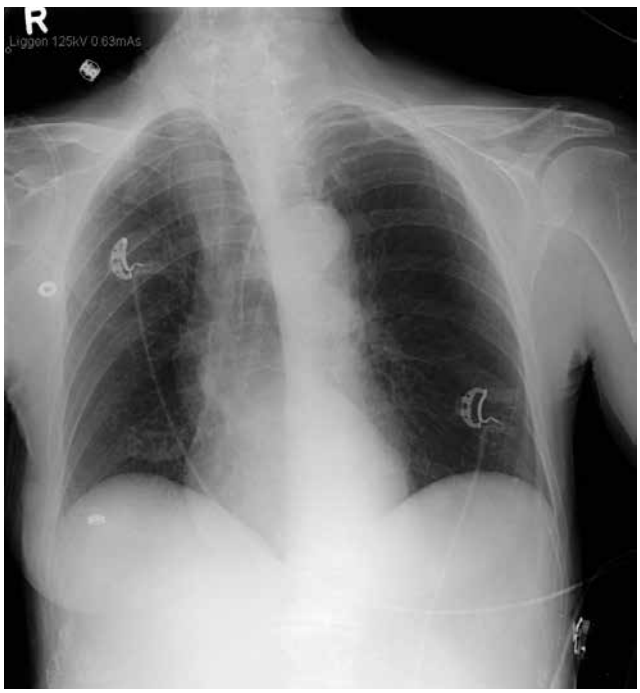
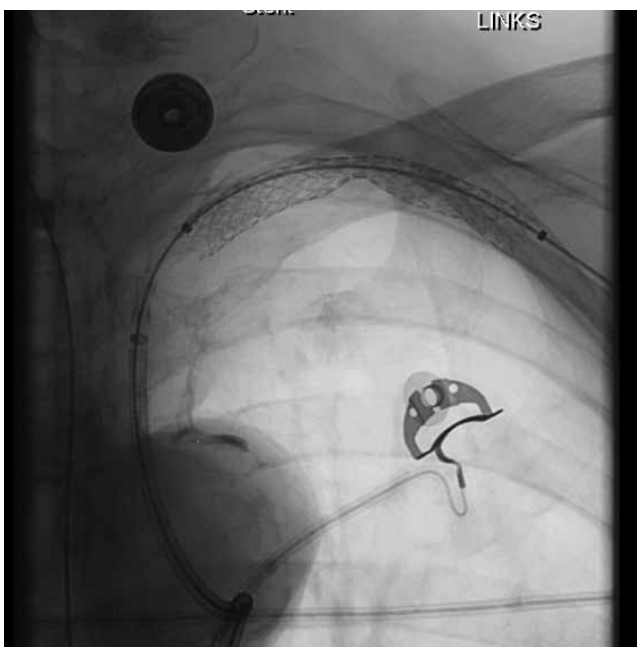


Figure 2. Stentgraft in correct position



19.

An outbreak of *Scabies norvegica* in the intensive care; a challenging diagnosis with severe consequences

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Background: In contrast to scabies vulgaris caused by *Sarcoptesscabiei mites*, variety hominis [1], crusted scabies or *Scabies norvegica* is a complicated and far more contagious form with a higher mite load, consequently making this type of scabies much easier to transmit.[1,2] Especially immunocompromised patients are vulnerable to this form of scabies.[2] If an intensive care unit is faced by this problem, adequate management is essential to control institutional spread.

Objectives: To raise awareness for the possibility of *Scabies norvegica*-infection in immunocompromised patients, which is – of course – often the case at an intensive care unit.

Methods: Descriptive case and overview of the measures taken to control and prevent this highly contagious infection.

Results: A 21-year-oldSomalian female with a known medical history of cystic fibrosis (CF) was admitted to our ICU with severe respiratory insufficiency, leading to a bilateral lung transplant one week later. Besides anticipated postoperative complications, a marked eosinophilia was present, for which an extensive differential diagnosis was investigated. Under the diagnosis of allograft rejection or an allergic reaction to medication (possibly morphine), she was treated with high-dose steroids. Despite this treatment, hypereosinophilia persisted. Also, she suffered from a pre-existing itch over her entire body, which persisted during further reconvalescence. Four months after admission she had developed multiple hyperkeratotic skin lesions. Dermatology diagnosed *Scabies norvegica*, confirmed by skin scrapings revealing several mites and eggs. Therapy with topical permethrin 5% and oral ivermectin was started, under which regimen her skin lesions resolved and the eosinophilia decreased. As a consequence, all patients close to and all of the healthcare workers involved in this patient's care were informed. A total of 185 healthcare workers were screened, while all symptomatic healthcare workers were treated with ivermectin. Furthermore, there was a significant economical burden, which was estimated at € 45,000, due to closure of ICU-beds and the costs of the comprehensive screening program.

Conclusions: In the majority of scabies-outbreaks, the index patient is an immunocompromised individual with unrecognized *Scabies norvegica*, who is more prone to develop crusted scabies because of masking of symptoms. Although rare, we therefore suggest that scabies be considered early in patients with itching skin-lesions of unknown origin, especially because of the burden and costs of the consequences of such an outbreak in terms of institutional precaution measures.

References

1. Choidow O. Scabies. New Engl J Med. 2006;354:1718-27.
2. Koene PRM, Tjioe M, Hoondert K, et al. Uitbraak van scabies in 1 ziekenhuis en 8 zorginstellingen door een geriatrische patiënt met scabiës crustosa. Ned Tijdschr Geneesk. 2006;150:918-23.

20.

Fungal infection causing ischemia of the spinal cord

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Case report: A 62-year-old woman diagnosed with acute myeloid leukemia was admitted to the intensive care unit with acute respiratory failure after being treated with the second induction cycle according to the HOVON 102B protocol. The chest-X-ray showed pleural effusion in the left lower quadrant while the laboratory results showed a leukocyte count of 0.1 x 10^9/L. The patient was intubated and mechanically ventilated. She was treated with filgrastim, vancomycin and meropenem for a suspected pulmonary infection. During the course of the treatment, she developed paraplegia of both legs with loss of sensation. An MRI of the spinal cord showed a paravertebral mass compressing the artery of Adamkiewicz, thereby causing spinal cord ischemia. A biopsy taken from this mass showed signs of a massive invasive fungal infection. The treatment was changed to liposomal amphotericin-B, to which the patient responded well. She was weaned from the ventilator and discharged from the intensive care unit, albeit with persistent paraplegia.

Discussion: Acute occlusion of the single artery of Adamkiewicz results in an anterior spinal artery syndrome that involves paraplegia with loss of reflexes, loss of pain and temperature sensation and loss of sphincter control with sparing of vibration and position sensation. The neurologic symptoms typically progress over a period of a few minutes to a few hours and recovery is poor.

Occlusion of the artery of Adamkiewicz has been described in a variety of conditions: preexisting abnormalities of the aorta, surgery of aorta, abdomen, spinal cord and lower limbs and spontaneous or traumatic thromboembolism. It has also been described in less common conditions such as epidural analgesia, cartilage embolus after a Valsalva maneuver, sclerotherapy for esophageal varices, and acute Schistosomamansoni infection. The main etiology of the anterior spinal artery syndrome during sepsis is compression of the anterior spinal artery by an epidural abscess, the most common causative organism being *S. aureus*.

Fungal infections causing paraplegia have been described only twice before. A patient with Crohn's disease treated with tumor necrosis factor antagonist therapy developed caseating granulomas in the spinal cord, revealing fungal hyphae consistent with Aspergillus and a patient with a prosthetic aortic valve experienced acute aortic occlusion by thrombotic material containing *Aspergillus niger*. Despite extensive antifungal therapy, both patients died. To our knowledge, this is the first report of an invasive fungal infection presenting as a mass

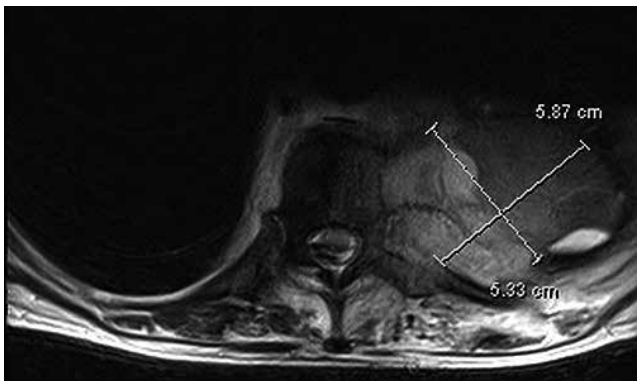
causing paraplegia by compressing the artery of Adamkiewizc, initially successfully treated with an antifungal agent. Voriconazole and liposomal amphotericin B are the antifungal agents recommended for the treatment of invasive fungal infections.[1] In addition, surgical resection may be curative, especially in case of persistent invasive aspergillosis and in patients needing additional immunosuppressive therapy.[2]

In conclusion, the case above illustrates that an invasive fungal infection may present as a mass compressing other structures, in this case leading to paraplegia due to spinal cord ischemia

References

1. Barberán J, Mensa J. Vallejo Llamas JC, et al. Recommendations for the treatment of invasive fungal infections caused by filamentous fungi in the haematological patient. Rev Esp Quimioter. 2011;24:263-70.
2. Danner BC, Didilis V, Dörge H, et al. Surgical treatment of pulmonary aspergillosis/mycosis in imuunocompromised patients. Interact Cardiovasc Thorac Surg. 2008;7:771-6.

Figure1. MRI (T2-axial): the lumbar spinal cord showing a paravertebral mass compressing the artery of Adamkiewicz



21.

Stone Heart Syndrome

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Introduction: Fourty years ago, severe ischemic contracture of the heart was a known, but uncommon complication of open-heart surgery using cardiopulmonary bypass. Since the introduction of myocardial hypothermia during surgery, this Stone Heart Syndrome is rarely seen anymore. However, to illustrate that this complication is not just a problem from the past, we describe a patient who recently underwent a standard mitral valve repair and developed a Stone Heart.

Case description: A 50-year-old man with severe mitral valve regurgitation, was admitted for a mitral valve repair. After starting cardiopulmonary bypass and before cross-clamping the aorta, a refractory ventricular tachycardia developed. The following mitral valve repair was uncomplicated. Immediately after discontinuing the perfusion, the left ventricle became thickened and hard as stone, the right ventricle contracted only slightly. There were no signs of local infarction or surgical complications. The electrocardiogram showed an asystole. A peripheral extracorporeal membrane oxygenation (ECMO) support system was implanted and the patient was transferred to a hospital with heart transplantation facilities. The next day the ECMO was centralised and blood and thrombi were removed from the pericardium. A large thrombus in the left atrium was left untouched because of the risk of air embolism. In the following days the heart did not recover and despite ECMO support he developed severe respiratory failure, acute kidney failure, liver failure and a persistent coma after discontinuing the sedation. Because of this multiple organ failure without therapeutic options, treatment was stopped nine days after the first operation. Autopsy revealed a heavy and solid heart, with thrombi in the left atrium, both ventricles and pulmonary veins and circular necrosis of the myocardium of both ventricles. Microscopy showed generalised coagulation necrosis and typical contraction bands in the myocardium. The coronary arteries were all open.

Discussion: The Stone Heart Syndrome was first described in 1972 and is typically occurring in patients undergoing open-heart surgery using cardiopulmonary bypass.[1]. During or after the extracorporeal perfusion, a sudden irreversible ischemic contracture of the heart develops. The hypothesis is that a combination of ATP depletion and intracellular calcium overload due to ischemia and reperfusion are responsible for the extreme contraction and the lack of relaxation.[2] Because of the contraction, intramyocardial coronary arteries are compressed, which causes secondary ischemia. Risk factors include left ventricular hypertrophy, aortic valve disease and pulmonary hypertension.[1] There are no therapeutic options, except for heart transplantation. Because of the contraction of the myocardium, no intraventricular assist device can be implanted. Therefore, the only way to bridge to decision is implanting an ECMO system. If the patient has no contraindications, the ECMO can bridge the patient to a heart transplantation. Induced hypothermia during surgery may prevent the Stone heart.[1]

This case illustrates that even today, patients without any risk factors undergoing a standard open-heart surgery procedure, may develop a Stone Heart Syndrome. If confronted with this phenomenon, an ECMO system may be implanted as a bridge to heart transplantation.

References

1. Cooley DA, Reul GJ, Wukasch DC. Ischemic contracture of the heart: "Stone Heart". Am J Cardiology. 1972;29:575-7.
2. Robinson RJ, Truong DT, Mulder D, Digerness SB, Kirklin JK. J Cardiothorac Anesth. 1989 Jun;3:361-8.