

**Report From Mongolia – How Much Do We Know About The Incidence Of Rare Cases  
In Less Developed Countries: A Case Series**

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## **ABSTRACT**

**Introduction:** Case reports are important instruments to describe rare disease conditions and give a rough estimation of their global incidence. Even though collected in international databases, most case reports are published by clinicians from industrialized nations and little is known about the incidence of rare cases in less developed countries, which are home to 75% of the world's population.

**Case Presentation:** We present seven patients who suffered from diseases which are either considered to be rare or have not yet been described before according to international databases, but occurred during a five months period in one intensive care unit of a less developed country. During the observation period, patients with a spontaneous infratentorial subdural hematoma (asian, female, 41 years), general exanthema and acute renal failure after diesel ingestion (asian, male, 30 years), transient cortical blindness complicating hepatic encephalopathy (asian, female, 49 years), Fournier gangrene complicating acute necrotizing pancreatitis (asian, male, 37 years), acute renal failure due to acetic acid intoxication (asian, male, 42 years), haemolytic uremic syndrome following septic abortion (asian, female, 45 years), and a metal needle as an unusual cause of chest pain (asian, male, 41 years) were treated. According to the current literature all seven disease conditions are considered either rare or have so far not yet been reported.

**Conclusion:** The global incidence of rare cases may be underestimated by contemporary international databases. Diseases which are currently considered to be rare in industrialized nations may occur at a higher frequency in less developed countries. Reasons may not only be a geographically different burden of certain diseases, limited diagnostic and therapeutic facilities, but also a relevant publication bias.

## **INTRODUCTION**

Case reports are important instruments to describe rare diseases and give a rough estimation of their global incidence [1]. Although collected in international databases, most case reports are published by clinicians from industrialized nations. Little is known about the incidence of rare cases in developing countries which are home to 75% of the world's population [2]. In this case presentation, we describe seven patients suffering from diseases which are either considered to be rare or have not yet been described before but occurred during five months in one intensive care unit (ICU) of a less developed country.

## CASES PRESENTATION

The eight-bed ICU is located in one of twelve university hospitals in the Mongolian capital of Ulaanbaatar and receives critically ill adult patients with surgical, medical and neurological pathologies. From July 1 until November 13, 2007, a total of 203 patients were treated.

Subsequently an overview of the clinical course of seven ICU patients with rare or so far unknown disease conditions each followed by a concise review of the current literature is presented. Table 1 summarizes demographic and clinical data of all patients. Written informed consent to anonymously present their history in this case presentation was obtained from all patients or their next of kin.

### *Patient 1 – Spontaneous Infratentorial Subdural Hematoma*

A 41 year old asian female presented to the emergency department with acute severe headache starting after a two days history of diarrhea. Cranial computertomography revealed an acute left-sided infratentorial subdural hematoma. A cerebral angiogram did not show any abnormalities. The patient denied recent trauma, intake of coagulation active drugs or herbs, or a known bleeding tendency. Plasma (prothrombin time, 10 sec; activated partial thromboplastin time, 28 sec) and cellular (platelet count, 165 G/L) coagulation parameters were normal. The patient was fully conscious but complained about nausea and vertigo. She was transferred to the ICU for neurologic monitoring and supportive therapy. Because of the non-compressive size of the hematoma, neurosurgical decompression was withheld. The patient was discharged from the ICU with significantly improved symptoms three days later.

Spontaneous subdural hematomas of the posterior fossa in adults without a history of trauma are very rare. Less than twenty cases have been reported in the literature [3]. Almost all were associated either with anticoagulation therapy or coagulatory defects.

### *Patient 2 – General Exanthema and Acute Renal Failure due to Diesel Ingestion*

During binge drinking a 30 year old asian male with a known allergy to diesel (local skin reactions) ingested an unknown amount of diesel ('several sips') when siphoning fuel off a canister. Within hours he developed fever, chills, coughing and general exanthema (Figure 1A and 1B). On day two, hematuria developed and progressed into oliguria. After seven days of cefazolin therapy because of pneumonia (Figure 1C) in a county hospital, the patient was admitted to the ICU with acute renal failure (creatinine, 740  $\mu\text{mol/L}$ ). Except for mild respiratory insufficiency and metabolic acidosis (pH, 7.29; standard bicarbonate, 13.5 mmol/L; base deficit, -12 mmol/L), he was stable. No history of cardiovascular instability could be evaluated. Liberate fluid resuscitation induced polyuria, decreased creatinine levels and evaded hemodialysis. Prednisolone was started at 80 mg and slowly tapered off after the general exanthema had improved.

Whereas localized dermatitis after diesel contact is known [3], no generalized exanthema following diesel ingestion has been reported. An allergic reaction seems to be the most probable cause in our patient. Acute renal failure has so far been observed in at least three patients after skin contact or diesel aspiration. Aliphatic hydrocarbons-induced hemolysis with hemoglobinuria, direct tubular toxicity, and allergic nephritis are possible pathogenetic mechanisms.

### *Patient 3 – Transient Cortical Blindness Complicating Hepatic Encephalopathy*

A 49 year old asian female with liver cirrhosis due to unspecified viral hepatitis was admitted to the ICU with coma (Glasgow Coma Scale, 9 pts; total bilirubin, 38  $\mu\text{mol/L}$ ; blood sugar, 7 mmol/L; arterial lactate, 2.5 mmol/L; plasma albumin, 35 mg/dL). Loss of consciousness was preceded by diarrhea followed by gradual visual impairment. Cranial computertomography, lumbar puncture and microbiological specimen were normal. In the electroencephalogram, triphasic waves and a delta rhythm were found. Two days after

initiation of enteral lactulose (6x30 mL/d) and supportive treatment, the patient's conscious state improved. Ophthalmologic examinations at ICU admission and after the patient had regained full consciousness revealed no ocular pathology. One week later, the patient was discharged from the ICU with improved but still impaired vision.

Cortical blindness associated with hepatic encephalopathy has first been described in 1979. Since then only four case reports have been published. As in our case, visual disturbances preceded the loss of consciousness in all reports. The pathogenesis of hepatic blindness is unknown but may include hypotensive episodes and impaired blood brain barrier function leading to cortical and subcortical edema [5].

#### *Patient 4 – Fournier Gangrene Complicating Acute Necrotizing Pancreatitis*

After binge drinking, a 37 year old asian man was admitted to the ICU with severe acute pancreatitis and multiple organ dysfunction (acute delirium; acute lung injury; total bilirubin 101  $\mu\text{mol/L}$ ; creatinine 238  $\mu\text{mol/L}$ ). His general condition improved with symptomatic ICU treatment. After a ten days course of antibiotic prophylaxis (4x1 g cefotaxime/day) pancreas necroses remained sterile (fine needle puncture). After ICU discharge, the patient developed extensive necroses of the skrotum and perineum (Fournier gangrene) requiring repeated surgical necrosectomy.

So far, scrotal involvement has been reported as a complication of acute necrotizing pancreatitis in four patients [6]. According to the current literature, necrosis culminating in Fournier gangrene is a yet unknown complication of pancreatitis. Comparable to the patients experiencing necrosis of the scrotum, descending retroperitoneal necroses most likely resulted in Fournier gangrene in our patient.

#### *Patient 5 – Acute Renal Failure due to Acetic Acid Intoxication*

During binge drinking a 42 year old asian chronic alcoholic male involuntarily ingested ~100 mL of 80% acetic acid. After hospital admission he developed intravascular hemolysis (hemoglobin, 57 g/L; lactate dehydrogenase, 3,752 IU/L) and acute renal failure (creatinine, 1,700  $\mu$ mol/L). When transferring the patient from the nephrological department to the ICU, he massively aspirated and died due to refractory pulmonary failure soon after ICU admission.

According to the experience of Mongolian physicians acetic acid ingestion is a frequent intoxication requiring hospital admission. At least three cases of acetic acid-associated acute renal failure are observed in the described hospital each year. In contrast, the current literature reports acute renal dysfunction to be a rare complication of acetic acid intoxication. So far six case reports/series have been published. As in the presented patient, hemolysis with hemoglobinuria caused kidney injury in most patients [7], but direct toxic effects of acetic acid on renal tubules may also be involved [8].

#### *Patient 6 – Hemolytic Uremic Syndrome Following Septic Abortion*

A 45 year old asian female suffered from septic abortion during gestational week 23. After curettage and initiation of antibiotic therapy (4x1 g ampicilline/d because of *E.Coli* growing from an intrauterine swap), the patient was stable and free of organ dysfunctions. On postoperative day two jaundice and oliguria developed, and she was transferred to the ICU. Despite of fluid resuscitation anuric renal failure (creatinine, 470  $\mu$ mol/L) developed. Repeated transfusions of red blood cells were required because of hemolytic anemia (hemoglobin, 61 g/L; lactatedeyhydrogenase, 3,027 IU/L). The blood analysis revealed fragmentocytes and thrombopenia (platelets, 48 G/L). Intermittent hemodialysis was started. One week after ICU admission, the patient was discharged with rising erythrocyte and platelet counts. Because of persistent anuria, hemodialysis was continued for another two weeks. Subsequently, renal function gradually returned to normal.

Although extraintestinal causes of hemolytic uremic syndrome are known [9], only one case following septic abortion has been published [10]. Comparable to intestinal hemolytic uremic syndrome, the *E.Coli* isolated from the uterine cavity in our patient did not only cause abortion but most probably also hemolysis and acute renal failure. Although further laboratory specification of the pathogen was not possible, it is likely that the clinical condition was caused by Shiga toxin [9].

#### *Patient 7 – A Metal Needle as an Unusual Cause of Chest Pain*

A 41 year old asian male presented to the emergency department with subacute recurrent chest pain. The electrocardiogram and biochemical laboratory parameters were normal. Chest fluoroscopy revealed a metal needle in the mediastinum. The patient could not remember of having swallowed the needle. Since no part of the needle could be reached through endoscopy, the patient was scheduled for surgery. Choosing a left lateral thoracotomy, the needle could only partly be removed because scar tissue most probably grew through the eye of the needle and prevented it from extraction without causing damage to the ventricular wall (Figure 2). Adhesions between the posterior pericardium and the esophagus suggested that the needle penetrated into the heart from the esophagus. The immediate postoperative course was complicated by respiratory problems but was uneventful afterwards. Three months after surgery, the patient still complained about intermittent mild chest pain.

Even though more than 100 cases of aorto-esophageal fistulas have been reported, only few cases of foreign body penetration into the heart are known [11].

## DISCUSSION

According to the literature all disease conditions presented are either considered rare (Patients 1, 3, 5, 6, 7) or have not yet been reported (Patients 2, 4). Although none of the illnesses was observed more than once the occurrence of seven such cases during a comparatively short time suggests an unusual accumulation, at least in the ICU evaluated. It is definitely possible that this is a finding by chance but it may also indicate that rare cases occur more frequently in less developed countries. This brings up the question if the global incidence of certain disease conditions is underestimated by the current literature which is largely based on reports from highly developed countries. It is at least worthwhile to reflect on why certain diseases assumed to be rare could occur more often in less developed countries.

The higher disease burden in developing countries [12,13] makes the occurrence of unusual cases more likely. Moreover, certain diseases (*e.g.* tropical diseases) are more frequent in the developing world. For example, the high incidence of chronic liver diseases in Mongolia [14] makes it probable that rare complications such as transient blindness associated with hepatic encephalopathy are observed more frequently. The widespread availability of potential toxins (*e.g.* acetic acid) and the lack of adequate protective measures (*e.g.* when handling fuel) results in a higher incidence of intoxications in less developed countries. Rare complications are further facilitated by variable diagnostic and therapeutic standards. It may be argued that earlier and more aggressive fluid resuscitation could have prevented acute renal failure in the two patients with acetic acid and diesel ingestion. Similarly, better imaging techniques and the possibility to perform interventional radiological procedures would have allowed earlier detection of the descending necroses and drainage in the pancreatitis patients.

Considering the low number of active scientists in developing countries [15], it is less likely that rare cases occurring in these regions are published. This may be the reason why

international databases suggest acute renal failure to be a rare complication of acetic acid poisoning while Mongolian physicians encounter this condition quite frequently. It may be hypothesized that further diseases remain either unknown or their global incidence underestimated simply because cases from less developed countries do not appear in international databases.

However, these points must not lead to the assumption that medical conditions which appear unclear in developing countries are rare cases that have not been observed in the medical literature. In contrast, it is much more likely that inadequate diagnostic facilities and limited educational standards preclude the diagnosis of well-known diseases. Given the possibility to perform appropriate diagnostic procedures and the clear disease presentation in our patients, it is unlikely that the presented cases are indeed “overlooked” common disease processes.

## **CONCLUSION**

The global incidence of rare cases may be underestimated by contemporary international databases. Diseases which are currently considered to be rare in industrialized nations may occur at a higher frequency in less developed countries. Reasons may not only be a geographically different burden of certain diseases, limited diagnostic and therapeutic facilities but also a relevant publication bias.

## **LIST OF ABBREVIATIONS USED**

ICU            intensive care unit

## **CONSENT**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal. In case of patient 5 who died consent for publication was sought from his next of kin.

## **COMPETING INTERESTS**

The authors declare that they have no competing interests.

## **AUTHORS' CONTRIBUTIONS**

MWD made substantial contribution to conception and design, interpreted the data and drafted the manuscript. OB gathered the data, interpreted the data and helped in drafting the manuscript. AHR gathered the data and helped in drafting the manuscript. WRH interpreted the data and helped in drafting the manuscript. GT made substantial contribution to conception and design, interpreted the data and helped in drafting the manuscript. All authors read and approved the final manuscript.

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## **FIGURE LEGENDS**

**Figure 1.** General exanthema (A, B) and left lower lobe pneumonia (C) after diesel ingestion.

**Figure 2.** Intraoperative situs during the procedure to extract a metal needle which sticks in the ventricular wall and caused subacute chest pain.

**Table 1.** Characteristics of Patients.

| <b>Patient</b> | <b>ICU Admission Diagnosis</b> | <b>Gender</b> | <b>Age</b> | <b>Chronic Disease</b>              | <b>SAPS II [60]</b> | <b>TISS 28 [61]</b> | <b>ICU LOS</b> | <b>ICU Outcome</b> |
|----------------|--------------------------------|---------------|------------|-------------------------------------|---------------------|---------------------|----------------|--------------------|
| 1              | Acute Infratentorial SDH       | F             | 41         | none                                | 10                  | 13                  | 3              | survived           |
| 2              | Intoxication with Diesel       | M             | 30         | allergy to diesel                   | 26                  | 18                  | 8              | survived           |
| 3              | Hepatic Encephalopathy         | F             | 49         | liver cirrhosis (Child A)           | 32                  | 15                  | 5              | survived           |
| 4              | Acute Necrotizing Pancreatitis | M             | 37         | chronic pancreatitis, alcohol abuse | 49                  | 28                  | 10             | survived           |
| 5              | Intoxication with Acetic Acid  | M             | 42         | alcohol abuse                       | 56                  | 30                  | 1              | died               |
| 6              | HUS after Septic Abortion      | F             | 45         | alcohol abuse                       | 38                  | 20                  | 7              | survived           |
| 7              | Foreign Body Extraction        | M             | 40         | none                                | 7                   | 18                  | 3              | survived           |

ICU, intensive care unit; SAPS, simplified acute physiology score; TISS, therapeutic intervention severity score; LOS, length of stay; SDH, subdural hematoma; HUS, hemolytic uremic syndrome.



