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McArdle's disease: case report of a rare condition in pregnancy

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Introduction

- McArdle's disease (glycogen storage disease type V) is an inherited disorder of glycogen metabolism affecting only skeletal muscle[1]
- A case of a pregnant woman with McArdle's disease is described here

Case report

Pre-op assessment

- 25 year old, nulliparous
- McArdle's disease- diagnosed at 16 years of age
- Symptoms- muscle cramps, tiredness on walking
- CVS- stable, normal echo
- Pregnancy- uncomplicated
- GA in childhood- uneventful

Anaesthetic plan

- In labour- early epidural advised for pain relief
- If operative delivery- regional anaesthesia preferred
- If GA necessary- anaesthetic preparations and management to mimic those for patients with malignant hyperthermia (MH)

Intrapartum management

- Spontaneous onset of labour at 41 weeks
- Early epidural sited for labour analgesia
- Delivered vaginally
- Blood loss 1200mL due to second degree perineal tear
- 1 unit of packed red blood cells transfused
- No complications at 24 hours postpartum

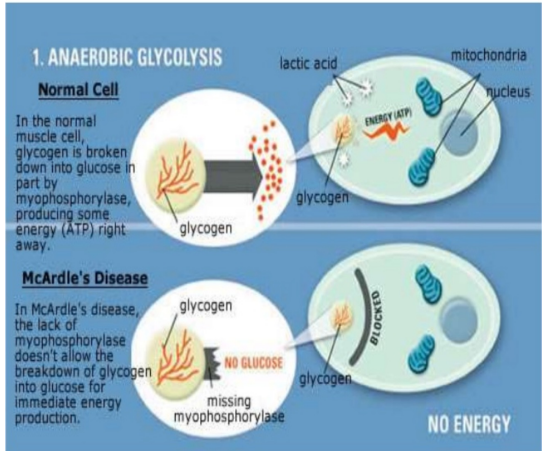


Figure1: Biochemistry of McArdle's disease

Discussion

McArdle's disease

- Caused by muscle glycogen phosphorylase deficiency resulting in inability to mobilise glycogen stores (figure 1)
- Skeletal muscle disorder, smooth muscle not affected
- Autosomal recessive inheritance, incidence of 1 in 100,000
- Symptoms include fatigue, muscle stiffness, myalgia and weakness which is induced by stress & exercise and relieved by rest
- No specific treatment, managed symptomatically
- Pregnancy and childbirth not adversely affected[2]

Anaesthetic implications

- Patients are at risk of hypoglycaemia, rhabdomyolysis, myoglobinuria and acute renal failure due to stress of surgery and anaesthesia
- Careful attention to be paid to fluid management, thermoregulation, maintenance of normoglycemia and avoidance of tourniquet
- Weak evidence of association between McArdle's and malignant hyperthermia[3]
- Malignant hyperthermia trigger substances to be avoided

Considerations during delivery

- Regional anaesthesia preferred as it decreases the likelihood that a woman with MH susceptibility will require GA
- Epidural analgesia recommended to reduce physical stress of labour with benefit of providing anaesthesia for emergency operative delivery
- Assisted delivery to shorten second stage of labour is beneficial

References

1. Quinlivan R, Buckley J, James M et al McArdle disease: a clinical review. Journal of Neurology, Neurosurgery & Psychiatry 2010;81:1182-1188

2. Giles W, Maher C. Myophosphorylase deficiency (McArdle disease) in a patient with normal pregnancy and normal pregnancy outcome. Obstet Med. 2011 Sep;4(3):120-1

3. Bollig G. McArdle's disease (glycogen storage disease type V) and anesthesia--a case report and review of the literature. Paediatr Anaesth. 2013 Sep;23(9):817-23