individuals with defective hepatic glucose-6-phosphatase, an essential enzyme in glucose homeostasis, who suffer with profound fasting hypoglycaemia. Clinical manifestations of the disease include hyperlipidaemia, renal disease and hepatocellular adenomas (HCA). In such patients, hepatic adenoma complications, or severely impaired metabolic control, are uncommon but recognised indications for liver transplantation.

We present the case of a 27-year-old woman with GSD-1a, who suffered with a plethora of secondary complications. Despite a complex dietary regime and extensive adjuvant medical treatment, her metabolic derangements remained resistant to treatment. She was initially referred for liver transplant due to recurrent episodes of life-threatening pancreatitis (secondary to hyperlipidaemia). Ultimately her multiple hepatic adenomas were accepted as indication for transplant. Due to the nature of her indication, her transplant priority was low, and she received an Orthotopic Liver Transplant (OLT) 3 years after referral. Although the adenomas appeared stable on regular pre-transplant MRIs, her explant sample showed histological evidence of hepatocellular carcinoma. Post-transplant, this patient's glucose metabolism and coagulopathy has resolved. She was initially treated for acute cellular rejection with good results. Her renal function remains stable and she reports a much improved quality of life.

Both the European and American College of Medical Genetics (ACMG) GSD1 guidance mentions OLT as a therapeutic option for adenomas suspicious of malignant transformation. This case highlights the shortfalls of using possible HCA transformation as a principle OLT indication. Serum markers are proven to be unreliable in patients with GSD1, and both MRI and CT are poorly predictive of transformation. In waiting for clinical features to develop as suggested by the ACMG, such as rapid enlargement or haemorrhage, patients risk dissemination of disease. The success of OLT in this patient suggests that severely poor metabolic control should be a more prominent recommendation for transplant in GSD1a.

There is evidence detailing post-liver transplant complications in GSD1a, with particular concern over post-operative renal function. Contrasting papers report promising long term outcomes. Due to the rarity of this condition, guidance is based on a handful of small scale case series. Therefore, it is important to document cases such as this one to contribute to GSD1a literature.

REFERENCES

1. Froissart R, Piraud M, Boudjemline AM, Vianey-Saban C, Petit F, Hubert-Buron A, et al. Glucose-6-phosphatase deficiency. *Orphanet J Rare Dis* 2011;**6**:27.
2. Rake JP, Visser G, Labrune P, Leonard JV, Ullrich K, Smit GPA. Guidelines for management of glycogen storage disease type I – European study on glycogen storage. *Eur J Pediatr* 2002 Oct;**161**(Suppl 1):S112–9.
3. Kishnani P, Austin S, Abdenur J, et al. Diagnosis and management of glycogen storage disease type I: a practice guideline of the American college of medical genetics and genomics. *Genet Med* 2014;**16**:e1.
4. Conti JA, Kemeny N. Type Ia glycogenosis associated with hepatocellular carcinoma. *Cancer* 1992;**69**:1320–1322.
5. Lee P. Glycogen storage disease type I: pathophysiology of liver adenomas. *Eur J Pediatr* 2002;**161**:S46–S49.
6. Labrune P. Glycogen storage disease type I: indications for liver and/or kidney transplantation. *Eur J Pediatr* 2002;**161**:S53–S55.

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DECOMPENSATED CIRRHOSIS CARE BUNDLE QUALITY IMPROVEMENT PROJECT IN LANCASHIRE TEACHING HOSPITALS NHS FOUNDATION TRUST

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The British Society of Gastroenterology (BSG) in collaboration with the British Association of the Study for Liver disease (BASL) produced a bundle to guide the management of decompensated cirrhotic patients(1) in the first 24 hours, aiming to improve mortality and reduce hospital stays in the United Kingdom. A variation of this guidance was adopted in Lancashire Teaching Hospitals NHS trust(LTHTR).

The aim of this study is to assess the compliance of LTHTR against the decompensated liver cirrhosis care bundle. Retrospective audit performed between December 2019 till March 2020 using data collected from electronic medical records. According to the presentation of decompensation, the management data was plotted against the guidance in the bundle.

The audit identified 35 patients with decompensated cirrhosis. The mean age of patients involved in the audit was 56 ± 27 , with Male patients(n =20) more than females (n=15). Some patients had more than one feature of decompensation on admission(n=15). Hospital stays ranged from 3 to 35 days with average of 12.5 days. Ascites was the major presentation of the patients included in this audit(n=19). However, only 5 patients had an ascitic tap done within 8 hours from senior review and 3 had antibiotics. 15 patients had hepatic encephalopathy(severity of which was not mentioned). Majority of those(n=11) received lactulose. Also, only 8 patients had documentation of Computed tomography (CT) heads considered or dismissed. Hyponatraemia or AKI was the 3rd commonest presentation within the audit's cohort(n=11). 9 patients had a medication review to stop possible drugs leading to AKI. However, only 6 of the patients had resuscitation fluids given on admission. Only 1 patient was weighed on admission, however, 8 patients were monitored for their input and output. Patients who presented with GI bleeds(n=6) had appropriately received platelets and Vit K when needed. Meanwhile, 4 of those patients received prophylactic antibiotics and only 1 received Terlipressin. Sepsis was the least common presentation throughout the audit cohort(n=4). However only 1 had a blood culture done. Out of all the patients that were involved in the audit 12 did not receive Venous Thromboembolism prophylaxis, when it was indicated.

The purpose of such a bundle was to reduce overall hospital stay and all-cause mortality of such patients. The findings were discussed in the local governance meeting which resulted in updating the bundle, establishing teaching sessions for doctors involved in patient management, updating the online system with an easy-to-follow flowchart to manage such patients. A second cycle will follow after implementation

Patient details

Decompensated Cirrhosis Care Bundle - First 24 Hours

Decompensated cirrhosis is a medical emergency with a high mortality. Effective early interventions can save lives and reduce hospital stay. This checklist should be completed for all patients admitted with decompensated cirrhosis within the first 6 hours of admission.

1. Investigations								
a)	NEWS ☐	FBC ☐	U/E ☐	LFT ☐	Coag ☐	Gluc ☐	Ca/PO ₄ /Mg ☐	
b)	Blood cultures ☐			Urine Dip/MSU ☐	CXR ☐	Request USS abdo ☐	CRP ☐	
c)	Perform ascitic tap in all patients with ascites using green needle irrespective of clotting parameters and send for ascitic PMN/WCC, culture and fluid albumin						Done Y N	N/A ☐
d)	Record recent daily alcohol intake				 Units			
2. Alcohol - if the patient has a history of current excess alcohol consumption (>8 units/day Males or >6 units/day Females) N/A ☐								
a)	Give IV Pabrinex (2 pairs of vials three times daily)					Y N		
b)	Commence CIWA score if evidence of alcohol withdrawal					Y N N/A		
3. Infections - if sepsis or infection is suspected N/A ☐								
a)	What was the suspected source?							
b)	Treat with antibiotics in accordance with Trust protocol					Y N		
c)	If the ascitic neutrophils >0.25 x 10 ⁹ /L (>250/mm ³)(i.e. SBP) then give:					Y N		
i)	Treat with antibiotics as per trust protocol					Y N NA		
ii)	IV albumin (20% Human Albumin solution) 1.5g/kg (20g of albumin in 100ml of 20% Human Albumin Solution)					Y N NA		
4. Acute kidney injury and/or hyponatraemia (Na <125 mmol/L) N/A ☐								
AKI defined by modified RIFLE criteria		1: Increase in serum creatinine ≥ 26μmol/L within 48hrs or						
		2: ≥50% rise in serum creatinine over the last 7 days or						
		3: Urine output (UO) <0.5mls/kg/hr for more than 6 hrs based on dry weight or						
		4: Clinically dehydrated						
a)	Suspend all diuretics and nephrotoxic drugs					Y N NA		
b)	Fluid resuscitate with 5% Human Albumin Solution or 0.9% Sodium Chloride (250ml boluses with regular reassessment: 1-2L will correct most losses)					Y N		
c)	Initiate fluid balance chart/daily weights					Y N		
d)	Aim for MAP>80mmHg to achieve UO>0.5ml/kg/hr based on dry weight					Y N		
e)	At 6 hrs, if target not achieved or EWS worsening then consider escalation to higher level of care					Y N NA		
5. GI bleeding – if the patient has evidence of GI bleeding and varices are suspected N/A ☐								
a)	Fluid resuscitate according to BP, pulse and venous pressure (aim MAP >65 mmHg)					Y N		
b)	Prescribe IV terlipressin 2mg four times daily (caution if known ischaemic heart disease or peripheral vascular disease; perform ECG in >65yrs)					Y N NA		
c)	Prescribe prophylactic antibiotics as per Trust protocol (cefuroxime unless contraindicated)					Y N		
d)	If prothrombin time (PT) prolonged give IV vitamin K 10mg stat					Y N NA		
e)	If PT> 20 seconds (or INR >2.0) – give FFP (2-4 units)					Y N NA		
f)	If platelets <50 – give IV platelets					Y N NA		
g)	Transfuse blood if Hb <7.0g/L or massive bleeding (aim for Hb >8g/L)					Y N NA		
h)	Early endoscopy after resuscitation (ideally within 12 hours)					Y N		

Continues overleaf..→

Please place in medical notes

Initials:

Time:

Initials:

Time:

Initials:

Time:

Initials:

Time:

Initials:

Time:

6. Encephalopathy			N/A ☐
a)	Look for precipitant (GI bleed, constipation, dehydration, sepsis etc.)	Y	N
b)	Encephalopathy – lactulose 20-30ml QDS or phosphate enema (aiming for 2 soft stools/day)	Y	N
c)	If in clinical doubt in a confused patient request CT head to exclude subdural haematoma	Y	N N/A
7. Other			
a)	Venous thromboembolism prophylaxis – prescribe prophylactic LMWH (patients with liver disease are at a high risk of thromboembolism even with a prolonged prothrombin time; withhold if patient is actively bleeding or platelets <50)	Y	N NA
b)	GI/Liver review at earliest opportunity (ideally within 24 hrs)	☐	

Initials:

Time:

Initials:

Time:

Name.....Grade.....Date.....Time.....

Decompensated Cirrhosis Care Bundle - First 24 Hours

The recent NCEPOD report 2013 on alcohol related liver disease highlighted that the management of some patients admitted with decompensated cirrhosis in the UK was suboptimal. Admission with decompensated cirrhosis is a common medical presentation and carries a high mortality (10-20% in hospital mortality). Early intervention with evidence-based treatments for patients with the complications of cirrhosis can save lives. This checklist aims to provide a guide to help ensure that the necessary early investigations are completed in a timely manner and appropriate treatments are given at the earliest opportunity.

- Decompensated cirrhosis is defined as a patient with cirrhosis who presents with an acute deterioration in liver function that can manifest with the following symptoms:
 - Jaundice
 - Increasing ascites
 - Hepatic encephalopathy
 - Renal impairment
 - GI bleeding
 - Signs of sepsis/hypovolaemia
- Frequently there is a precipitant that leads to the decompensation of cirrhosis. Common causes are:
 - GI bleeding (variceal and non-variceal)
 - Infection/sepsis (spontaneous bacterial peritonitis, urine, chest, cholangitis etc)
 - Alcoholic hepatitis
 - Acute portal vein thrombosis
 - Development of hepatocellular carcinoma
 - Drugs (Alcohol, opiates, NSAIDs etc)
 - Ischaemic liver injury (sepsis or hypotension)
 - Dehydration
 - Constipation

When assessing patients who present with decompensated cirrhosis please look for the precipitating causes and treat accordingly. The checklist shown overleaf gives a guide on the necessary investigations and early management of these patients admitted with decompensated cirrhosis and should be completed on all patients who present with this condition. The checklist is designed to optimize a patient's management in the first 24 hours when specialist liver/gastro input might not be available. Please arrange for a review of the patient by the gastro/liver team at the earliest opportunity. Escalation of care to higher level should be considered in patients not responding to treatment when reviewed after 6 hours, particularly in those with first presentation and those with good underlying performance status prior to the recent illness.

Stuart McPherson, Jessica Dyson, Andrew Austin, Mark Hudson 2014

Abstract P088 Figure 1