Development of AIH care bundle

Jess Dyson

Freeman Hospital

Committee

- Jess Dyson (chair)
- Andrew Yeoman (doctor)
- Neil Halliday (doctor)
- Lin Lee Wong (doctor)
- Mussarat Rahim (doctor)
- Sarah Flatley (doctor)
- Yooyun Chung (doctor)
- Ann Brownlee (patient representative)
- Megan Morgan (specialist nurse)
- Irene Salala (specialist nurse)

Background

- Rare disease – 17 per 100,000
- Heterogeneous presentation and disease course
- Challenging diagnosis and histology
- Complex cases
- Significant morbidity and mortality
  - Disease and it’s treatments
- Need to standardise and optimise care

Are we meeting our treatment goals?

UK-AIH

- Obtain biochemical remission
  - 59% in biochemical remission
- Aim for azathioprine (or MMF) monotherapy
  - 29 different treatment regimens
  - 55% on long-term steroids
- Prevent progression/transplant
  - Reduced survival
    - 10-40% become cirrhotic despite treatment
    - ~7% of adult elective OLT in UK
- Improve patient quality of life
  - Lower QoL from disease and treatments

Hoeroldt et al.Gastroenterol 2011. Feld et al. Hepatol 2005. Roberts et al. Gastroenterol 1996. Verma et al. Am J Gastroenterol 2004.Czaja et al.Hepatol 2006.Gleeson and Heneghan.Gut 2011.Schramm et al.J Hepatol 2013.Sockalingam et al.J Hepatol. 2012

Patient name:
NHS number:

Primary Biliary Cholangitis (PBC) Care Bundle

This checklist is designed to aid your patient's management and guide escalation decisions. Suitability for treatment and surveillance may be considered according to performance status and comorbidity.
Guidance for all domains can be found overleaf.

Please ensure the patient has been signposted to appropriate support groups.

Treat & Risk Stratify				
Treated with UDCA?		Y	N	Intol
UDCA dosed at 13-15mg/kg?		Y	N	N/A
Eligible for second-line therapy?		Y	N	
Treated with second-line therapy?		Y	N	N/A
Actively Manage				
Assessed for symptoms?	Pruritus	Y	N	
	Fatigue	Y	N	
	Sicca	Y	N	
Osteoporotic fracture risk assessed in the last 5 years?		Y	N	
Stage & Survey				
Disease staged with non-invasive test in last 3 years?		Y	N	N/A
If cirrhotic: surveillance imaging within the last 6 months?		Y	N	N/A
If clinically significant portal hypertension: variceal screening in date (or takes NSBB)?		Y	N	N/A
If evidence of decompensation or bilirubin > 50 µmol/L: discussed with transplant centre?		Y	N	N/A
Additional Comments				

First-line Treatment

- Ursodeoxycholic acid (UDCA) dosed at 13-15mg/kg/day

Second-line Treatment

- Review indications for second-line treatment (SLT) at every clinic appointment. If SLT is indicated, refer to local SLT MDT* for consideration of obeticholic acid and/or fibrates.
  **referral not required in Scotland*

Indications
- Intolerance of UDCA
- Inadequate biochemical response to UDCA (after 12 months of treatment at the optimum dose), defined as:
  - **alkaline phosphatase (ALP) > 1.67 x ULN** and/or
  - **bilirubin > 1 x ULN**

Contraindications
- Child-Pugh B or C cirrhosis
- Child-Pugh A cirrhosis with portal hypertension

Bone Health

- Use risk assessment tools e.g. FRAX or QFracture
- For patients with a clinically significant risk of fracture ensure appropriate action as per NICE guidelines: https://cks.nice.org.uk/topics/osteoporosis-prevention-of-fragility-fractures/

Cirrhosis

- Stage with non-invasive tools e.g., elastography, Enhanced Liver Fibrosis (ELF) score
- Ensure 6-monthly USS for HCC surveillance if cirrhotic
- If evidence of clinically significant portal hypertension commence varices surveillance in keeping with national cirrhosis and portal hypertension guidelines
- If evidence of decompensation (ascites, hepatic encephalopathy, variceal haemorrhage, bilirubin ≥50 µmol/L) discuss with liver transplant centre.

Pruritus:

Supportive therapy – to be used at all stages
Aqueous cream / E45 PRN
Menthol 1% in aqueous cream PRN
Antihistamines for relief of night-time itching

First-line

Bile acid sequestrants (e.g., Colestyramine 4g OD, titrate to max of 16g/day, Colesevalam 625mg BD, titrate to 3.75g/day)

Give 2-4hrs before or after other medications

If no improvement after 4 weeks at maximum tolerated dose, stop and move onto second-line therapy.

Second-line

Rifampicin: 150mg OD, titrate to max 600mg/day

Requires LFT monitoring

Check for drug interactions prior to use (including hormonal contraceptives and hormonal replacement therapy)

Try for 8 weeks at maximum tolerated dose.

If pruritus not controlled with second-line therapy, **seek specialist hepatology opinion**

Fatigue:

Treat direct contributors

- Ensure pruritus is treated to minimise sleep disturbance
- Screen for associated autoimmune disease (e.g., AIH, thyroid disease, B12 deficiency, Addison's disease)
- Check for age-related conditions (e.g., diabetes, heart failure, renal failure)

Modify exacerbating factors

- Screen for depression
- Screen for autonomic dysfunction
- Assess sleep hygiene

Lifestyle adjustments

- Recommend pacing strategies (using available energy to its best advantage) and timing strategies (arranging key tasks for earlier in the day).

Support

- Signpost patients to support groups (e.g., PBC Foundation, British Liver Trust)

Sicca symptoms:

Dry eyes: artificial tears, lubricating eye ointment (night-time)
Dry mouth: good dental hygiene, sugar-free chewing gum and lozenges, artificial saliva
For refractory symptoms refer to specialist management (rheumatology/ophthalmology)

Link to full BSG PBC Guidelines : https://www.bsg.org.uk/wp-content/uploads/2019/11/BSG-and-UKPBC-primary-biliary-cholangitis-treatment-and-management-guidelines.pdf

Concept of bundle

- Streamlined as possible
- Usability
  - non-specialists
  - specialist nurses
  - trainees
- Ideal minimum data set
- Key metrics
  - biochemical remission
  - need for treatment escalation or potential reduction
  - symptoms/side effects
  - complications

Bundle structure

1. Clinical history summary
   how we got to where we are

2. Clinic review
   where we are

3. Guidance notes

1. Clinical history summary

- Diagnosis
- Overlap?
- Disease stage
- Mode of presentation
- TPMT

IAIHG Diagnostic Criteria

Feature/parameter	Discriminator	Score
Antibodies (max 2)		(0–2 total)
ANA or SMA+	≥1:40	+1
ANA or SMA+	≥1:80	+2
or LKM+	≥1:40	+2
or SLA/LP+	Any titre	+2
IgG or γ-globulins level	>ULN	+1
	>1.1x ULN	+2
Liver histology (evidence of hepatitis is required)	Compatible with AIH	+1
	Typical of AIH	+2
	Atypical	0
Absence of viral hepatitis	No	0
	Yes	+2

Typical biopsy features
- interface hepatitis
- plasma cells
- rosetting
- emperipolesis

Score ≥7 = Definite AIH
Score ≥6 = Probable AIH

Hennes EM, et al. Hepatology 2008;48:169–76; EASL CPG AIH. J Hepatol 2015;63:971–1004

Diagnostic Details

Date of diagnosis		Month:	Year:
Immunology at diagnosis	ANA	Pos/neg/not done	Titre
	SMA	Pos/neg/not done	Titre
	LKM	Pos/neg/not done	Titre
	SLA	Pos/neg/not done	Titre
	LC1	Pos/neg/not done	Titre
	AMA	Pos/neg/not done	Titre
	M2	Pos/neg/not done	Titre
	IgG	Value:	
Diagnostic liver biopsy	Month/year:	Result:	
	If no biopsy, why not:		
Details of further biopsies	Month/year:	Result:	
Evidence of overlap syndrome	Yes/No	Details:	
Cirrhotic at diagnosis	Yes/No	Evidence for cirrhosis:	
Mode of presentation	Acute liver failure Acute hepatitis with jaundice Acute hepatitis without jaundice Incidental finding of abnormal LFTs Other (give details):		
TPMT result	High/low/normal/not done		

1. Clinical history summary

- Previous immunosuppressive treatment(s)
- Other important health conditions
- Screening for common associated disorders
  - thyroid, haematinics, vit D, coeliac

2. Clinic review - treatment

Current weight:

Shared care agreement with primary care: Yes/No

Current treatment

If not on treatment, why:

Medication	Total daily dose	Weight based (mg/kg, where relevant)	Date last given
Prednisolone			
Budesonide			
Azathioprine			
Mercaptopurine			
Allopurinol			
Mycophenolate mofetil			
Mycophenolic acid			
Tacrolimus			
Rituximab			
Other	What and dose:		

Remission status

Complete biochemical remission (normal ALT and IgG) Yes/No

Date achieved biochemical remission:

Reason for not targeting 'normal':

2. Clinic review – holistic care

- Holistic care
  - Mood
  - Quality of life
  - Treatment side effects
  - Treatment adherence
  - Vaccinations
  - Pre-conception planning/contraception (where relevant)
  - Steroid Emergency Card and sick day rules
  - Lifestyle advice

2. Clinic review – staging and surveillance

All patients			
Bloods (inc HbA1c if on steroids)	What tests:	Frequency:	
Osteoporosis (https://cks.nice.org.uk/topics/osteoporosis-prevention-of-fragility-fractures/)	FRAX score	Date:	Result:
	DEXA	Date:	Result:
	Vitamin D level	Date:	Result:
	Vitamin D indicated	Yes/No	
	Bisphosphonate indicated	Yes/No	
Non-cirrhotic patients (non-invasive assessment every 2-3 years)	Fibroscan	Date:	Result:
	Ultrasound	Date:	Result:
Cirrhotic patients - 6 monthly HCC USS	Date:	Result:	
Cirrhotic patients – portal hypertension	OGD	Date:	Result:
	NSBB	Yes/No	

2. Clinic review – review and F/U

Review and Follow-Up Plan

Interval to next bloods		
Interval to next clinic/which clinic		
Tests requested	Fibroscan	Yes/No/Not needed
	Ultrasound	Yes/No/Not needed
	DEXA	Yes/No/Not needed
	OGD	Yes/No/Not needed

3. Guidance notes

- Triggers to consider referral to specialist centre
- Goals of therapy
  - complete biochemical remission (CBR)
    - Adherence, metabolites, review diagnosis (esp DILI and viruses)
- Review risks and benefits of treatment at each appointment
- Avoid budesonide in cirrhosis
- Pregnancy
- Young adults/transition
- Signpost/provide patient support

Autotext/smart codes

- Vaccinations
  - *Please note*: All adult patients on immunosuppression are advised to have the seasonal flu, COVID, pneumococcal and non-live shingles vaccine (e.g. Shingrix) vaccines. Please can you ensure that this is offered to the patient via your practice. Live vaccines (e.g. Yellow fever, MMR) should be avoided in some immunosuppressed patients. Please contact the hepatology team or consult the Greenbook guidance if necessary
- Sick day rules
- Patient support

Potential pitfalls

- Balance between detail and usability
- Clinical history summary takes a bit of time
- User engagement
- Different care delivery models

Current progress

- Very much in work in progress
- Testing current bundle in clinic
- Honest feedback

The future

- Integrate into electronic patient record
  - EPIC
  - Potential for data extraction

- Patient facing version
  - how to prepare for your consultation
  - list of questions and prioritise your questions

- Endorsement from BSG/BASL

Taking Stock of AIH in the UK

Wed 11th February 0930-1700
Porter Hall, The Wesley Euston, London

5 CPD points

Contact: lmoore@bsg.org.uk

bsg

BASL
British Association for the Study of the Liver

UK-AIH

Thank you

Any comments or questions?