APPROACH TO A CHILD WITH CHRONIC HEPATITIS

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Hepatology symposium,
Definition of Chronic liver disease

- progressive process

- End point is liver parenchymal destruction, fibrosis and cirrhosis

- usually time dependent—6 months
12 yr Female

- Fatigue,
- Tinge of Jaundice
- Hepatomegaly–2cm, Splenomegaly +
- Bilirubin 50/30 um/l
- AST/ALT 540/630 uL
Other tests

- INR – 4.1
- Total protein: 100g/
- HB 10
- WBC – 2800
- Platelets – 130,000
- Ultrasound – Non – homogenous nodular liver; enlarged spleen.
Synthesis of case

- Teenage girl
- Severe liver disease, chronic (US)
- Mild jaundice
- Portal hypertension: Spleen (hypersplenism)
- Liver failure (or vitamin K deficiency?): PT
- Increased protein ↔ gammaglobulins?
Other tests

- **Gammaglobulins**: 32 g/l
  - ➔ Autoimmune disease or cirrhosis

- **Auto antibodies**
  - antinuclear + : 1/400
  - anti-smooth muscle + : 1/640

  ➔ autoimmune hepatitis

- You learn later aunt has thyroiditis and cousin has diabetis: you should have asked!
HYPOTHESIS ?

- **Virus** : B vaccinated, C negative, D
  - ( A neg, but no chronic disease and no liver failure without jaundice)

- **Genetic**
  - Wilson, alpha-1-AT deficiency, cystic fibrosis, Hereditary haemachromatosis, glycogen storage disease

- **Autoimmune**
  - Autoimmune – Hepatitis, sclerosing cholangitis,
  - Primary biliary cirrhosis
Non-alcoholic fatty liver disease (NALFD)
- DM related, obesity, metabolic syndrome

Drugs
- INH, methotrexate, valproate, nitrofurantoin

Vascular
- Veno-occlusive conditions - Budd Chiari

Conditions from infancy manifesting later - PFIC, storage diseases, CF, choledocal cyst

Idiopathic/cryptogenic
CLINICAL APPROACH

Principles

- Confirm presence of CLD
- Assess severity
- Etiological diagnosis
SUGGESTIVE CLINICAL FEATURES

- Past history of conjugated hyperbilirubinaemia in infancy
- Family history of chronic liver disease, genetic or autoimmunity
- Recurrence in apparent acute hepatitis
- Persistence of clinical features of acute hepatitis beyond 3 months
- Previous history of HBV, HCV or non A non E
- Drugs ingestion
Liver—small, enlarged left lobe, nodular
Splenomegaly, Ascites, Oedema
cutaneous portosystemic shunts
Portal hypertension
Growth failure
Cutaneous signs—telangiectasia, spider angioma, finger clubbing etc
Endocrine signs—testicular atrophy, rickets
Extrahepatic autoimmune manifestations—
Kayser fleischer rings
Baseline work up

- FBC—Normocytic–chromic anaemia, +/- leucopenia, thrombocytopenia
- LFTs—deranged, transaminitis, could be normal in advanced disease. INR, PT–
- Urea+ electrolytes—hepatorenal syndrome or iatrogenic effect(diuretic tx)

NOTE:
PT/INR, Albumin, platelets—tests of synthetic liver function. Derangement=severe disease
VIRAL STUDIES
– HBsAg, HBeAg, anti-Hbe, HBV DNA
– anti–HCV ab, HCV PCR
– Liver biopsy

WILSONS DISEASE
– Slit lamp exam–KF rings
– caeruloplasmin, plasma copper
– urinary copper–baseline + post penicillamine challenge
– Neuro exam + MRI
– Liver biopsy–fat, mallory bodies etc, copper density
Wilson disease showing copper deposition in zone 1 hepatocytes. Rhodanine (Emanuele-Goodman modification).
- High transaminitis and IgG
- +VE ANA, ASMA, ANCA in AIH type 1 and sclerosing cholangitis.
- +LKM –1 in AIH type 2
- Magnetic resonance /retrograde cholangiogram–abnormal in ASC
- Liver biopsy–interface hepatitis and multilobular collapse–hallmark
ALPHA 1–ANTIPRYSIN DEFICIENCY
–alpha-1 antitypsin serum levels
–PiZZ phenotype by isoelectric focusing
–Liver biopsy– characteristic globules on PAS

CYSTIC FIBROSIS
–Sweat test, genetic testing, lung function tests

PROGRESSIVE INTRAHEPATIC FAMILIAL CHOLESTASIS
–Low GGT in PFIC 1, high GGT in MDR3 def

STORAGE DISEASE
–nieman pick, glycogen storage disorder– genetic testing
CHRONIC OUTFLOW OBSTRUCTION
– Ultrasound hepatic and portal veins flow
– Echocardiogram
– Cardiac catheterization

MASS LESIONS
– Choledochal cyst, liver tumours
– Ultrasound
Principles of treatment

Supportive–

- Role of diet–only restrict proteins if in liver failure otherwise ensure balanced diet
- Mx of cholestasis– ADEK, MCT, Choleretics, Antipruritic agents, dietary modifications
- Antibacterial agents, antifungals in liver failure.
- Adequate rest, hydration and avoidance of further insults to liver eg hepatotoxic medications
Definitive treatments

- Specific antiviral therapy for hepatitis B/C, HIV

- Steroids and immunomodulating agents

- Replacement of the deficient abnormal end product, such as oral administration of primary bile acids (in patients who have abnormalities of bile acid biosynthesis)
Molecular manipulations (eg, inhibition of polymerization of alpha 1–antitrypsin)

Receptor–based, targeted enzyme replacement therapy

Gene therapy

Hepatocyte transplantation (eg, fulminant liver failure)

Liver transplantation
Chronic liver disease is common

Work up based on clinical presentation and knowledge of CLD etiology is more rewarding than blind work up
THANK YOU