UNRECOGNISED SCURVY WITH HOME PARENTERAL NUTRITION

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Introduction The classical dermatological signs of vitamin C deficiency are now rare. The diagnosis of Scurvy was initially unrecognised in a 16-year-old boy.

Aim To describe a challenging case of vitamin C deficiency.

Subject and Method A 16-year-old boy with intestinal failure due to short bowel syndrome. He had been on home parenteral nutrition (PN) for 14 years. He had restricted dietary intake due to intestinal rapid transit.

He presented with 4 weeks history of non-pruritic rash on his legs and 2 weeks of bruising with ankle and knee pain.

On examination, he was systemically well. There were numerous peri-follicular petechiae distributed symmetrically over his lower legs with a few scattered lesions on the arms and trunk. (see figures 1, 2).

Initial investigations did not reveal blood dyscrasia, infection, or autoimmune disorder. Vitamin A, D, E, B1, B12, folate, Zinc, Copper, Selenium, and Manganese were within normal limits.

Rheumatology and dermatology opinions were sought; diagnosis of Scurvy was suspected. Skin Biopsy showed perifolliculitis with erythrocyte extravasation suggestive of Scurvy.

Four days after starting vitamin C supplement, symptoms completely resolved. Subsequently, plasma vitamin C level came back low at 1.6 μmol/L (<11 μmol/L indicates deficiency).

Immediate contact was made with the compounding company and an enquiry revealed inadvertent use of a uni-layer bag for his home PN, allowing accelerated degradation despite addition of the appropriate amount at manufacture.

Summary and discussion PN is a lifesaving treatment for patients who cannot be adequately nourished by other feeding routes. Despite this, nutritional deficiencies still occur, warranting close nutritional monitoring.

Analysis of ascorbic acid in the uni-layer bag showed degradation by 30% every 24 hours due to oxidation and by 14 days virtually none was detected. Our patient used bags stored for up to 14 days. In a multi-layer bag, 50% degradation occurs over 14 days, but the amount remaining in the bag was above the recommended daily allowance of 100 mg/day. Rate of vitamin C decay depends on the constituents of PN, storage environment and the bag containing it (1, 2).

Conclusion This case is a reminder of the Scurvy’s clinical presentation. Diagnosis was initially challenging; however, starting treatment on clinical suspicion while awaiting lab results relieved the symptoms and substantiated the diagnosis.

Scurvy is preventable and may occur with well monitored PN support as well as in fully enterally fed children (3). We advise to ensure home PN bags are multi-layered and practice stock rotation not only when nearing the expiry date.

Note: Consent was obtained to publish the photos.

REFERENCES

UPTAKE OF NO-BIOPSY APPROACH IN DIAGNOSING PAEDIATRIC COELIAC DISEASE IN A REGIONAL TERTIARY CENTRE

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Background Coeliac disease (CD) is an immune mediated systemic disorder strongly associated with HLA DQ2 and DQ8 haplotypes.

In 2012 The European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) recommended serology based No-Biopsy Approach to the diagnosis of CD if the following criteria are met1:

REFERENCES
Serum levels of IgA antibodies against type–2 (tissue) transglutaminase (TGA–IgA) ≥ 10 times the upper limit of normal (≥10x ULN)
Positive endomysial antibodies (EMA–IgA) in a second serum sample
Positive coeliac HLA risk alleles DQ2 and/or DQ8
Symptoms suggestive of CD (particularly malabsorption)

These guidelines were modified in 2020 recommending:
HLA testing and presence of symptoms are not obligatory criteria for a serology based diagnosis without biopsies.

**Aim**
To review the practice in a Regional Paediatric Gastroenterology Centre on the uptake of No-Biopsy Approach to the diagnosis of CD comparing the 2012 and 2020 guidelines.

**Abstract P58 Figure 1**

**Total Patients with TGA-IgA ≥10 times ULN + Duodenal Biopsy (n=68)**

- TGA-IgA≥10 x ULN+EMA-IgA+ Symptoms+ HLA risk allele + duodenal biopsy= 19%
- TGA-IgA≥10 x ULN+EMA-IgA+ symptoms +duodenal biopsy =91%
- TGA-IgA≥10 x ULN+EMA-IgA +duodenal biopsy =95%
- TGA-IgA≥10 x ULN+duodenal biopsy = 100%

**Abstract P58 Figure 2**
Abstract P59 Figure 1  The t-test shows a statistically significant difference in the fluorescence values in normal and steatotic livers samples (p<0.01), t=3.55, df= 17

Abstract P59 Figure 2  A) Steatotic hepatocytes stained with nile red and DAPI. B) Background fluorescence in unstained steatotic hepatocytes. C) Non-fatty hepatocytes stained with nile red and DAPI. D) Background fluorescence in unstained non-fatty hepatocytes.

USE OF NILE RED TO ASSESS QUALITY OF STEATOTIC DONOR HUMAN HEPATOCYTES

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Introduction With the rise in obesity and insulin resistance, there is an increased incidence of liver diseases such as Non-Alcoholic Fatty Liver Disease (NAFLD) and Non-Alcoholic Steatotic Hepatitis (NASH), which can lead to the development of liver cancer. Liver biopsy is the gold standard for classification of liver steatosis (mild, moderate, or severe) to determine stage of NAFLD and NASH and the suitability of donor livers for transplantation. However, these observations are made visually and thus are prone to inter-observer variations.

Aim The aim of the project is to develop a simple quantitative test to assess fat content in isolated human hepatocytes.

Methods Nile Red was used to stain lipids in human hepatocyte batches (14 steatotic and 8 non-steatotic). 10,000 cells were placed per well in a black 96-well plate and diluted Nile Red solution was added. Plates were incubated (37°C, 10 min), then read using a fluorescence plate reader. Data were recorded and analysed. Cytospins of 10,000 cells per slide were allowed and the slides were then air-dried for 24h. They were stained with Nile Red (humidity chamber at 37°C, 10 min), cell nuclei were counterstained with DAPI, and slides were visualised under a fluorescence microscope. Images were taken. [In both techniques unstained cells were used as negative controls].

Results The t-test showed statistically significant difference in fluorescence values in steatotic vs non-steatotic hepatocytes.

REFERENCES

Design/Methods A 6-year retrospective study of all children who attended coeliac clinic at The Great North Children Hospital, Newcastle.

Data obtained using electronic patient records included TGA-IgA, EMA-IgA, symptoms at initial presentation and histopathological reports.

HLA typing results were obtained from the Regional NHS blood and transplant laboratory.

346 children with CD were reviewed in the coeliac clinic from July 2013 to July 2019. Age range 0.9 – 16.5 years (median 9.5 years) and 54% female.

Exclusion criteria include diagnosed outside study period or the UK, TGA-IgA at initial presentation unavailable for review.

Results 66% of cases had TGA-IgA ≥10xULN at initial presentation. 48% were diagnosed by serology based No-Biopsy Approach (figure1).

Duodenal biopsies were performed in 82 cases. Biopsies were performed for type1 diabetes -8.5% and asymptomatic patients with first-degree relative with CD -8.5% (figure 1).

EMA-IgA positivity was reported in 65/68 cases with symptoms attributed to CD in 62 cases in the cohort. A total of 13/62 cases had HLA risk alleles DQ2 and/or DQ8 performed (figure2).

Conclusion(s)
• HLA screening uptake was 63%. The low uptake of HLA typing may have contributed to the increased number of cases undergoing duodenal biopsies.
• Based on 2012 guidelines 19% of cases had duodenal biopsies for diagnosis of CD despite meeting the criteria for no biopsy approach.
• Based on 2020 guidelines 96% of cases had duodenal biopsies for diagnosis of CD despite meeting the criteria for no biopsy approach.
• The changes in the CD guidelines from 2012 to 2020 have resulted in an increase from 16% to 96% of cases that may have benefitted from no biopsy approach to the diagnosis of CD.
• A unifying approach to the diagnosis of the CD will reduce the variability in investigations.
• The current restrictions to Aerosol Generating Procedures due to SARS-CoV-2 pandemic will have a positive impact on establishing a No-Biopsy approach to the diagnosis of CD.