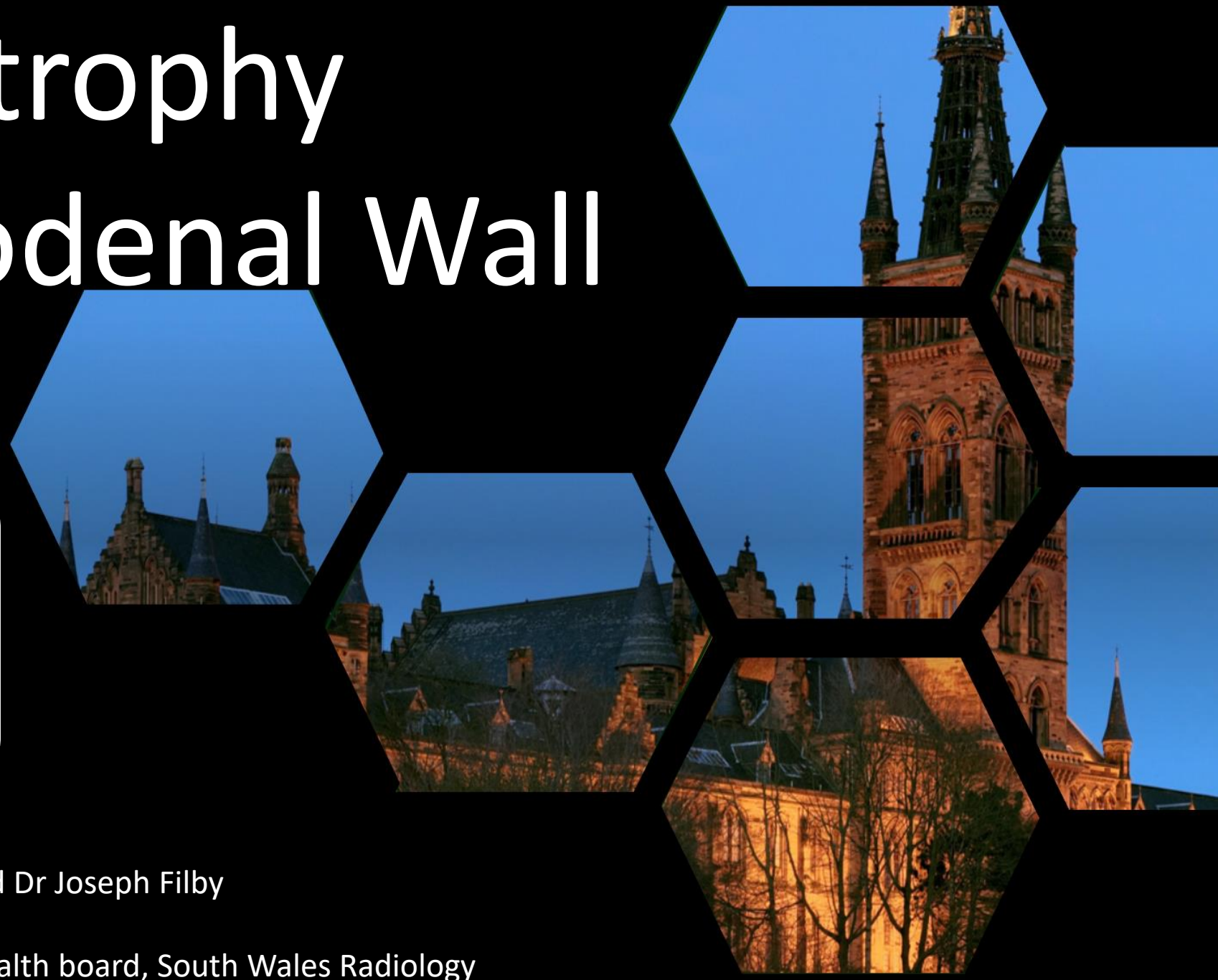
Cystic Dystrophy of the Duodenal Wall



A Case Report
and
Analysis of Current Literature

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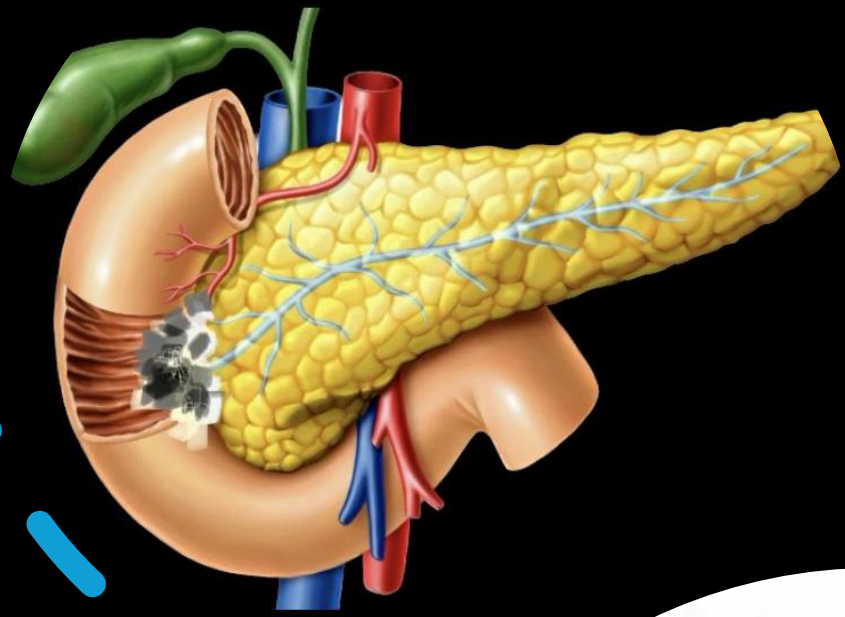
Aetiology

Cystic Dystrophy of the duodenal wall (CDDW) is thought to be due to inflammation of ectopic pancreatic tissue in the wall of the duodenum

The condition is characterised by mural cystic changes on imaging.

CDDW is part of a spectrum of pathologies that are grouped under the umbrella term “paraduodenal pancreatitis”[1].

Other entities which also fall under this umbrella term include, groove pancreatitis and myoadenomatosis.



Literature Review

- CDDW is a rare disease
- Only 64 papers were identified when a literature review was performed using keywords: *“cystic dystrophy of the duodenal wall"* and *“heterotopic pancreatic tissue in the duodenum”*
- Estimating a prevalence of 0.2-15% from post-mortem studies and case reports [2-6].
- 80-85% of cases have a history of alcohol abuse with a mean age of 45 at diagnosis[3-5]. CPPD has a male predominance.
- ~ 90% present with epigastric pain, 40% weight loss and 35-40% with vomiting [2-5].
- Only 42-45% of cases have deranged LFT and diagnostically elevated Amylase[5].

Risk Factors:

Risk factors for developing CDDW include:

- Smoking
- Long-term alcohol misuse
- History of previous episodes of pancreatitis [2].

Case Report

- We report the case of a 42-year-old male with a background of alcohol abuse.

- Over a 2-year period, he presented on 12 occasions to hospital complaining of epigastric pain, vomiting and unintentional weight loss.

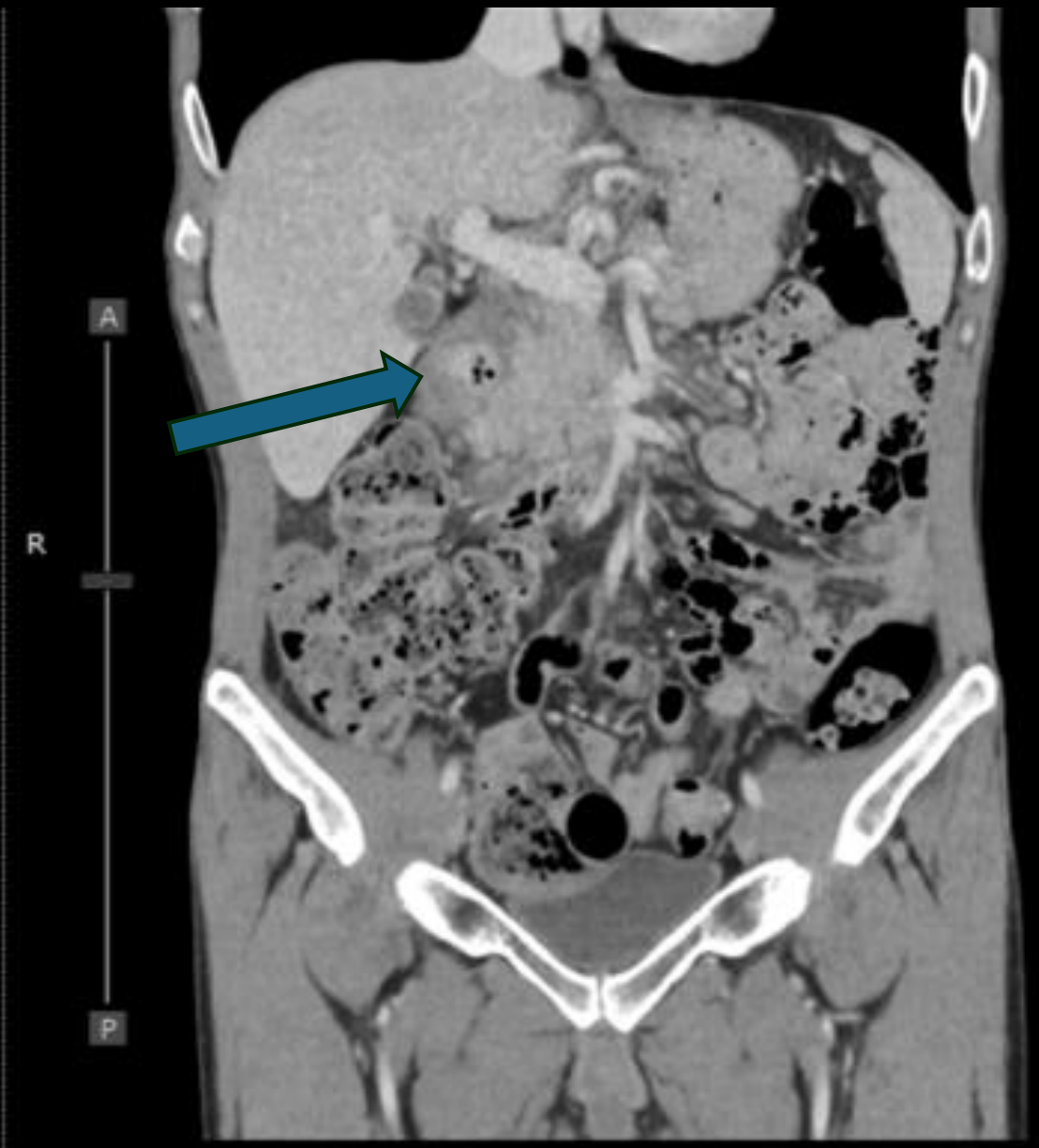


Figure 1. Coronal CT:
Fluid in the region of the D2 flexure and pancreatic head (blue arrow)

Initial Presentation

INITIAL PRESENTATION TO ACCIDENT AND EMERGENCY WAS IN AUGUST 2021 WITH EPIGASTRIC PAIN AND MILDLY ELEVATED CRP, WCC AND AMYLASE.

CT DEMONSTRATED PERI-GASTRIC AND PERI-DUODENAL FREE FLUID WHICH WAS MIS-DIAGNOSED AS A WALLED-OFF DUODENAL PERFORATION.

THE PATIENT WAS SUBSEQUENTLY MANAGED MEDICALLY WITH A LONG COURSE OF IV, THEN ORAL ANTIBIOTICS

Amylase Levels

Date and Time Collected	U/L	Min	Max
14-Jul-23 11:13	*109		100
13-Jul-23 11:20	68		100
08-Jun-23 08:25	56		100
06-Jun-23 15:01	100		100
02-Jun-23 00:00	*174		100
09-May-23 10:55	*120		100
07-May-23 08:28	72		100
27-Apr-23 16:41	*150		100
19-Apr-23 08:45	*133		100
07-Sep-22 08:10	*181		100
28-Apr-22 10:25	*179		100
13-Apr-22 00:00	*299		100
14-Mar-22 11:00	*182		100
19-Jan-22 12:44	*281		100
13-Oct-21 09:50	*201		100
30-Aug-21 13:00	*159		100

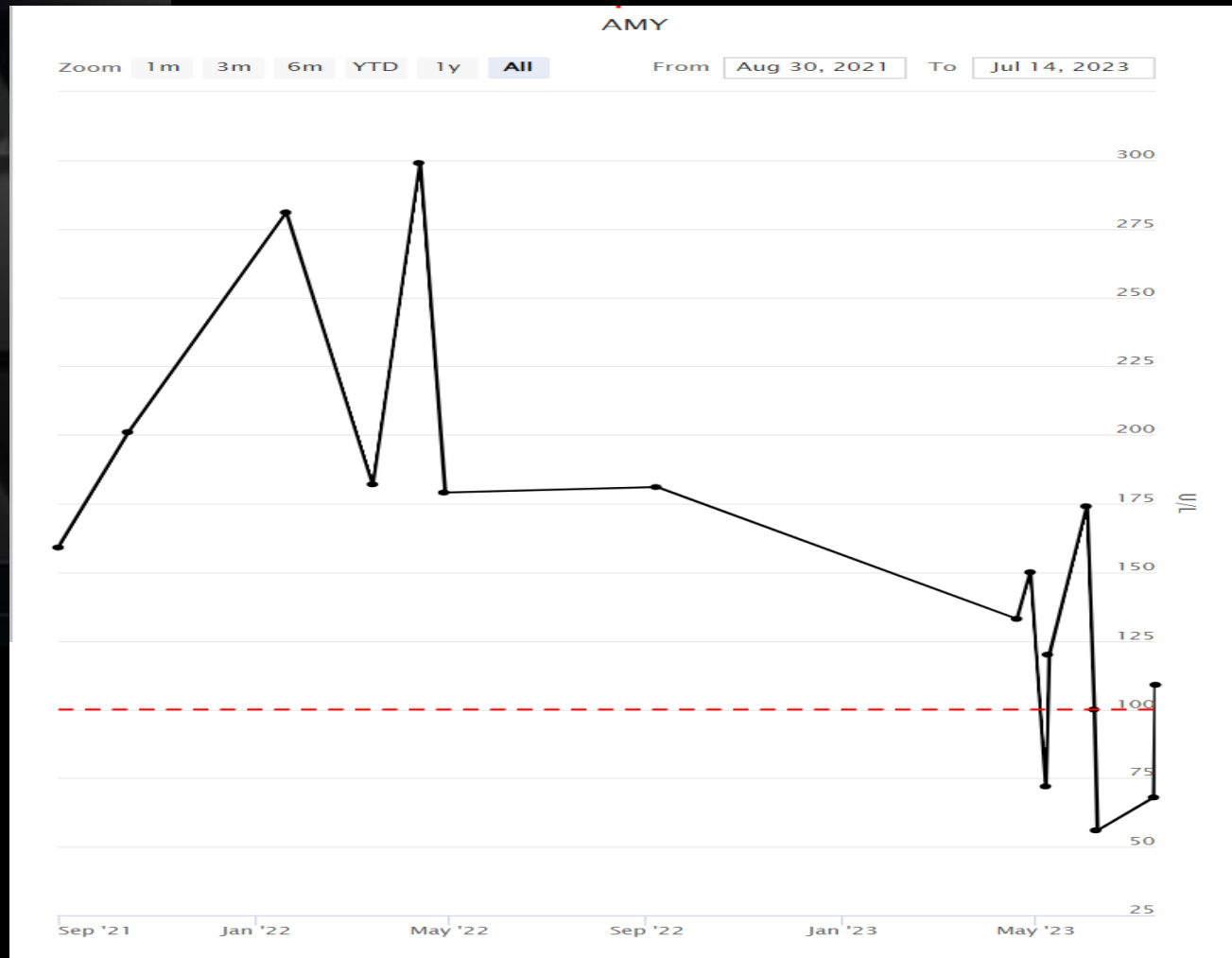
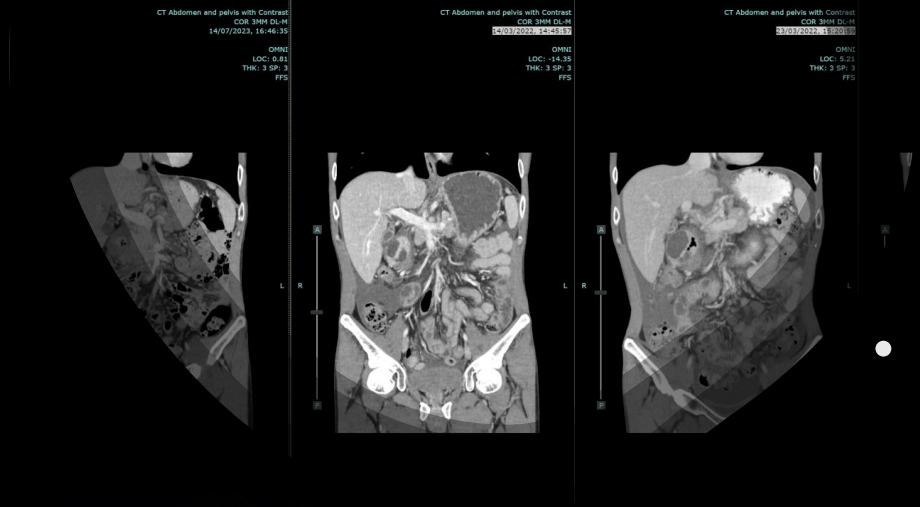


Figure 2. Graph:

Blood amylase levels (vertical axis) over a two-year period (horizontal axis) remained below diagnostic levels.



Repeated Admissions

- The patient had repeated admissions with a total of 12 hospital presentations for his symptoms of epigastric pain, vomiting and weight loss.

- He underwent a total of :
- Six abdominal CT studies
- Four transabdominal ultrasounds
- One water soluble contrast study
- Two oesophagogastroduodenoscopies (OGDs).

- Following radiology review CDDW was suspected.

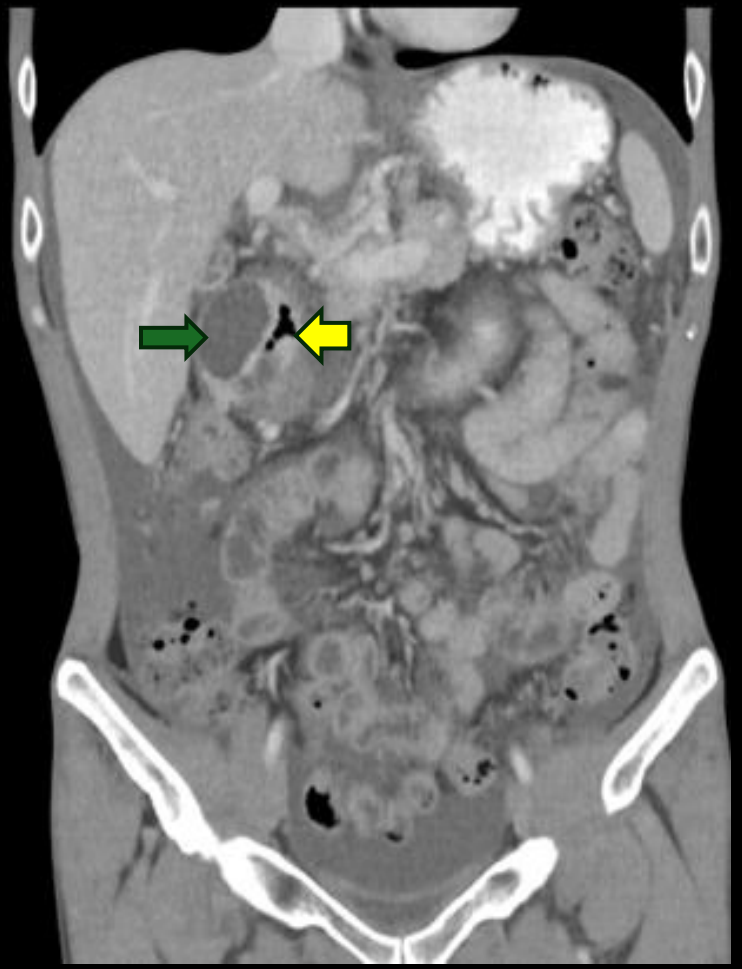


Figure 3. Coronal CT:

Thick walled, multiloculated cysts within the duodenal mucosa(green arrow), adjacent to the duodenal lumen (yellow arrow)

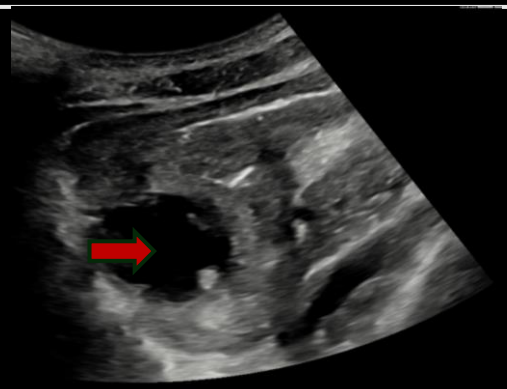


Figure 4. Trans-abdominal Ultrasound :

Low echoic cystic lesion in the region of the second part of the duodenum (red arrow).

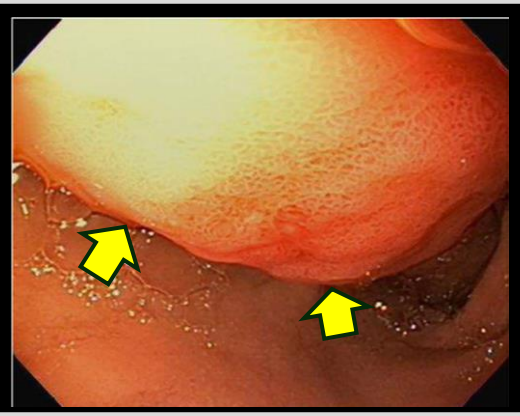


Figure 5. Endoscopy:

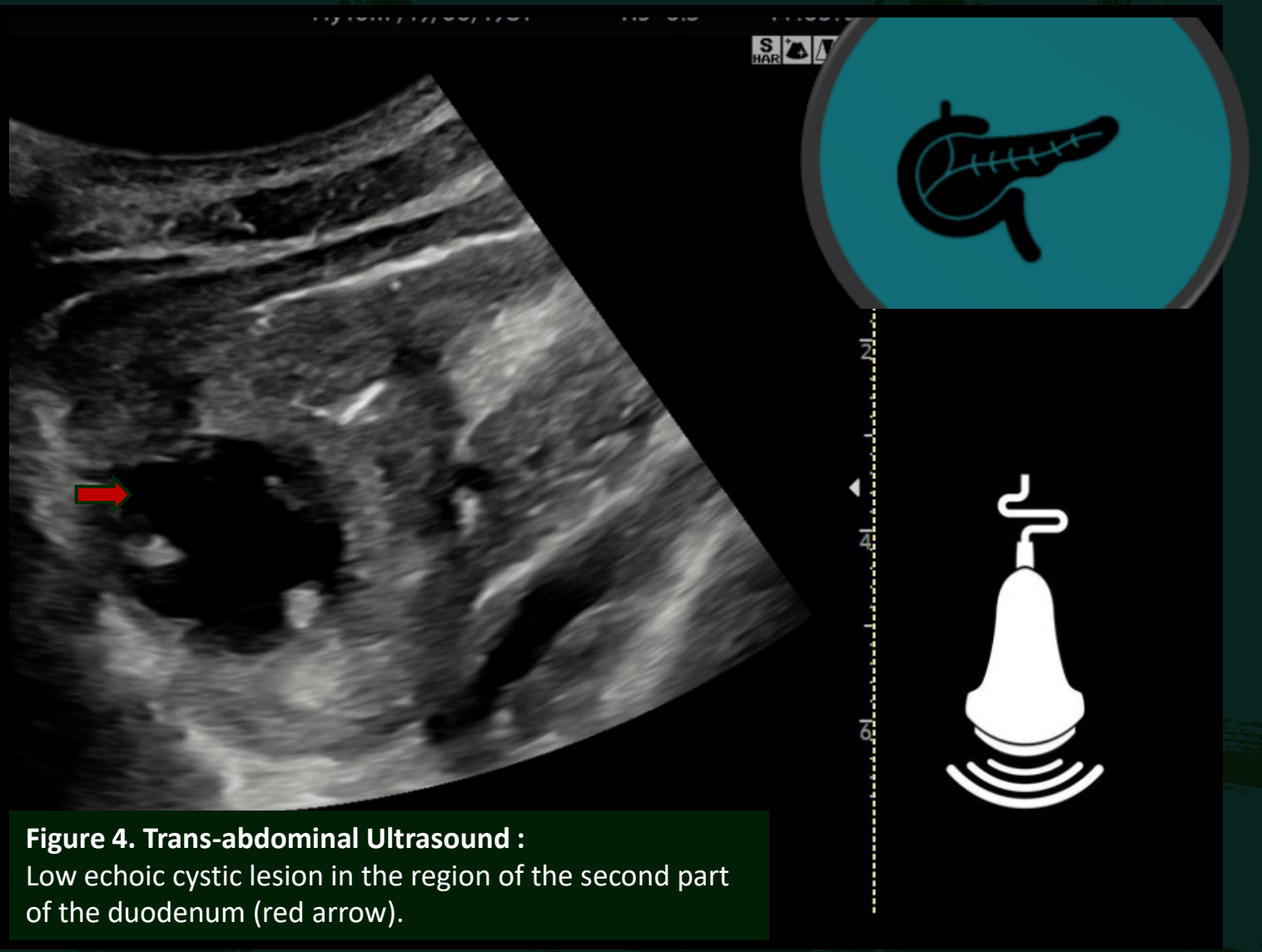
Mucosal out-pouching in the second part of the duodenum (yellow arrows) (7).

Investigations

Figure 3-5. Demonstrate investigations performed over the two-year period

Histology

- US- guided percutaneous aspiration of a duodenal cyst was undertaken.
- Subsequent analysis of the aspirate revealed amylase levels of 38,583 iu/L.
- This was consistent with a diagnosis of CDDW.



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Management

- The patient was referred to The Welsh Regional Pancreaticobiliary Surgery Centre */ Canolfan Ranbarthol Llaweddygaeth Pancreaticobiliary Cymbru*.
- For Consideration of Pylorus Preserving Pancreatoduodectomy (PPPD)
- **Outcome**: No malignancy identified and advised trial of continued medical management with PPI and alcohol abstinence.
- Planned for 6/12 review if symptoms remain unresolved patient would be for further consideration of PPPD.

References

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6. Campos L.P., Mateu C.A., García-Argüelles J.S., Durá Ayet A.B., Pérez I.B., Callol P.S.: Cystic dystrophy of the duodenal wall: a rare but need-to-know disease. Endosc Ultrasound 2017; 6: pp. 61-66

7. Endoscopy image taken from : Sarrazin B. D, Rao D. Thimmappa N. Cystic dystrophy of the duodenal wall: case report and literature review. Clinical Imaging, 2021-10-01, Volume 78, Pages 113-116.