In our subject the presence of macroamylasemia was excluded as (also -complic). The periodic control of oncological markers (CA19-9) was negative. In 2018, after 38 years from the first finding of hyperenzymemia, the patient performed an MRI of the upper abdomen with the following result: liver containing some small cystic formations scattered in both lobes, the greater than 10 mm; pancreas within limits; multiple bilateral renal cortical cysts, the largest of 22 mm; no areas of pathological enhancement after paramagnetic contrast medium infusion (gadodiamide) of the upper abdomen organs.

The magnetic resonance cholangiopancreatography (MRCP) showed no biliary lithiasis, no dilatation of the intrapancreatic biliary tree, hepato-choleodochus with a maximum caliber of 7 mm, regular Warsing duct (fig. 3.4 a).

The laboratory tests performed in February 2019 have demonstrated: total amylase 151 U/L (nv: 10-120), lipase 364 U/L (nv: 2-67), yGT 53 U/L (nv: < 55), blood glucose 107 mg/dL (nv: 60-100), total cholesterol 197 mg/dl, triglycerides 153 mg/dl. The most recent laboratory tests (September 2019) have shown: lipase 219 U/L (nv: 13-60), a-amylase 117 U/L (nv: 28-100), pancreatic iso-amylase 75.8 U/L (nv: 13-53), faecal calprotectin 81 ug/g faeces (nv: < 50). CA 19-9 18.80 KU/L (nv: < 3), positive occult blood in the stool. In October 2019, colonoscopy showed an ulcer in the right flexure; biopsy: adenok (right hemicolectomy). Abdomen CT demonstrated pancreas with initial aspects of adipose-kutobal trophie without expansive formations, multiple liver cysts with a diameter of less than 1 cm, to the kidneys some simple cortical cysts, the largest of 22 mm in diameter.

Conclusions.

1. Our clinical case meets the criteria for the definition of chronic asymptomatic pancreatic hyperenzymemia. In fact the inclusion criteria are: an increase by > 10% of the normal value of serum amylase and/or lipase found on >3 occasions, lasting more than 6 months, absence of upper abdominal or back pain; idopathic presentation.

2. We excluded the presence of any associated congenital anomalies (pancreas divisum, annular pancreas, Wurschungocele, cistic lesions at the pancreatic tail, Santorini, diffuse dilatation of the main pancreatic duct, intraductal papilary mucinous tumor of the pancreas), alterations that have been described in some cases (1). We did not perform a MRCP with secretin stimulation (S-MRPC), currently considered as the method of choice in the study of CAPH subjects (10), for the search for sphincter of Oddi dysfunction or even early (mild) chronic pancreaticosis: this investigation would not have added significant data to the already stable clinical picture. Similar considerations apply to the use of endoscopic ultrasonography (EUS) (1-6).

3. In our family group two brothers with hyperenzymemia are carriers, a third brother carries only hepatic and renal cysts. Of three siblings without hyperenzymemia, two carries multiple pancreatic, hepatic and kidney cysts, a third has died for complications (pancreas and breast) at the age of 79 years. High incidence (5 out of 10) of neoplasia in siblings. The significance of these associations at the present time is not clear.

4. Regarding the study of CFTR (cystic fibrosis transmembrane conductance regulator) gene mutations and SPINK1 and PRSS1 genes, no significant differences compared to the general population have been described in benign familial hyperenzymemia (7).

5. In our case the long follow-up (30-39 years) confirms the benignity of the enzymatic anomaly regarding pancreatic diseases.

6. The prevalence of CAPH among subjects who underwent blood tests for multiple pancreatic serum enzymes was 2% in a retrospective cross-sectional observational study in a large sample of the general Italian population (8).

7. The coexistence of hyperenzymemia in siblings is diagnostic for familial form of CAPH, form that has an incidence between 4% and 39% in the series described in the literature (9-11). This familial association supports the concept of a genetic basis underlying enzyme abnormalities, despite the fact that so far no correlations have been found with the genetic mutations studied.

References.