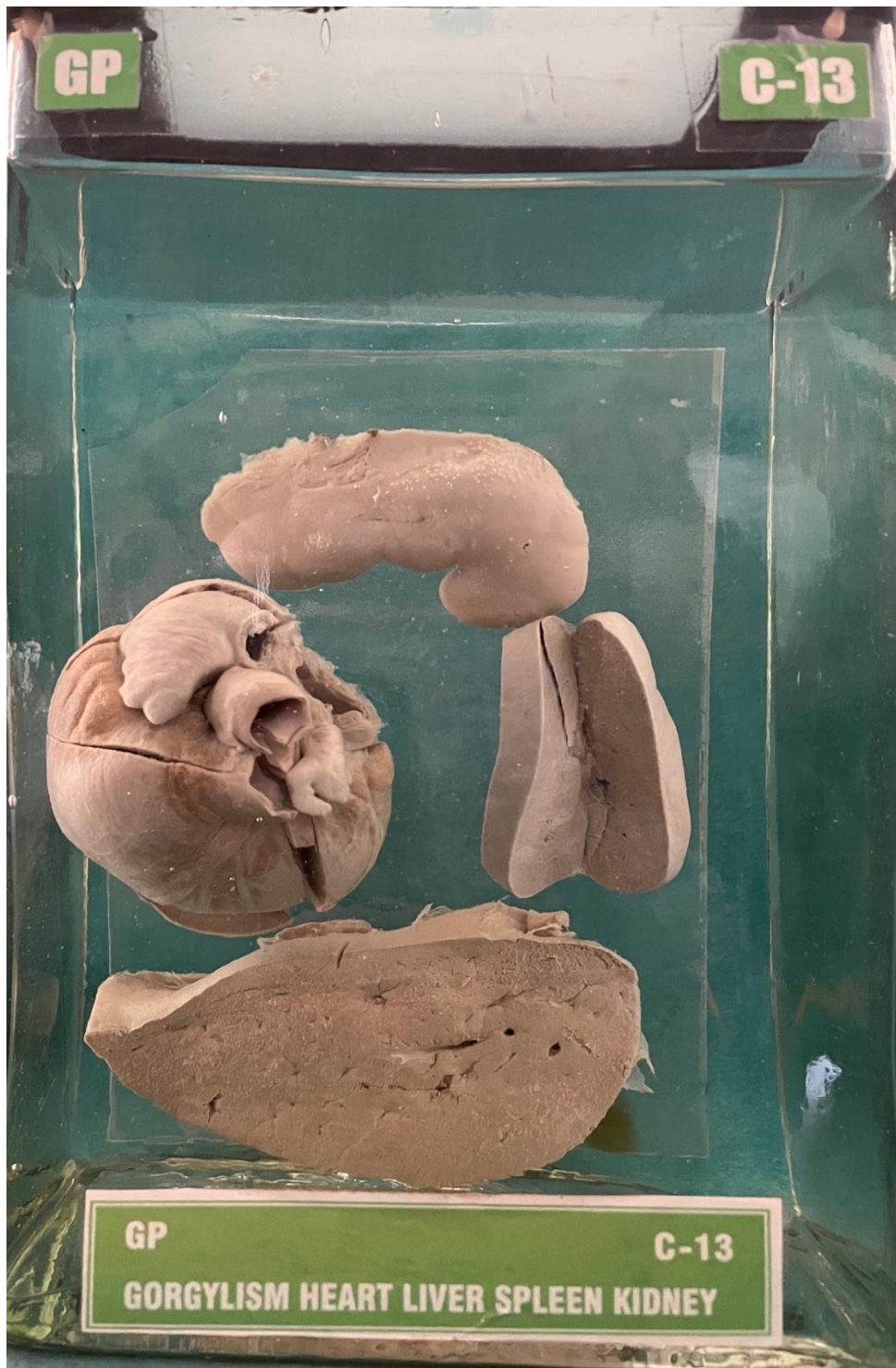


C13 – GORGOYLISM HEART LIVER SPLEEN KIDNEY



HISTORY:- A child aged 1 1/2 years, was admitted in the Civil Hospital Belgaum, with history of puffiness of face and abdominal pain and rigidity, since one month. Delayed mild stone. The case was clinically diagnosed as Hurler's syndrome. Patient expired after 4 days of admission. Clinical post mortem was conducted.

MACROSCOPY:- Specimen of heart. The heart is enlarged and globular. There is hypertrophy of myocardium. Valves appear normal.

Specimen of liver. The liver is enlarged. The outer surface is smooth. Cut section is yellowish white homogeneous in appearance.

? Specimen of spleen, enlarged in its size and has lobulated appearance.

Specimen of kidney. Outer surface is smooth. Cut section shows pale homogeneous appearance.

MICROSCOPY:- Sections from heart show normal structure. The myocardial cells in the subendocardial zone and just beneath the epicardium show vacuoles in the cytoplasm. The vacuoles are of varying sizes. Frozen section and special stains technic showed absence of fat.

Sections from spleen showed normal structure with trabeculae containing large irregular cells with vacuolated cytoplasm.

Sections from kidney show normal structure with tubular epithelium showing vacuoles in some places.

IMPRESSION:- Heart, liver, spleen, kidney in Gorgoylism.