Results:
- 7 females and 6 males
- Consanguineous marriages were reported in 11 patients (84.6%).
- Mean age at diagnosis: 4.4 years (0.5-17).
- Median follow-up period: 7.2 years (0.8-13)
- The beginning of neurodegeneration age: 16 months (min: 6, max: 72)
- One patient required tracheostomy and mechanical ventilation during the follow-up.
- All patients were treated with a low lysine diet and carnitine supplementation.

Background:
Glutaric acidemia type I (GA1) is an inherited neurometabolic disorder caused by deficiency of glutaryl-CoA dehydrogenase activity. Most untreated patients are asymptomatic at birth and then develop irreversible neurological impairment with encephalopathic crises.

Materials and Methods:
The clinical and laboratory findings from the last fifteen years were reviewed retrospectively.

Cranial MRI Findings:
- White matter changes (100%)
- Frontotemporal atrophy (92.3%)
- Basal ganglia (84.6%)
- Subarachnoid abnormalities (46.1%)
- Subdural hematoma (15.3%)

Laboratory:
- High glutaric acid (%100) (mean: 661±sd mmol/molKrea)
- High 3-hydroxyglutaric acid (%100) (mean:19.6 mmol/molKrea)

Discussion:
In our study, unlike classical knowledge, most patients did not have macrocephaly. The neurodevelopmental delay and neurodegeneration were observed in all cases.