**INTRODUCTION:** Glutaric Aciduria type 1 (GA-1) is produced by the deficiency of the enzyme glutaryl-CoA dehydrogenase, accumulating glutaric acid and 3-hydroxyglutaric acid (GA, 3-OH-GA). The diagnosis is made by Tandem Mass Spectrometry (MS/MS) and confirmed by analysis of urine organic acids (UOA). Patients without early treatment have an acute encephalopathic crisis (AEC), followed by irreversible neurological symptoms. The treatment consists of a diet restricted in lysine, special formula (SF), L-carnitine and riboflavin supplementation. **OBJECTIVE:** To present the chilean experience in diagnosis and follow-up of patients with GA-1. **METHODOLOGY:** 41 records of patients diagnosed with GA-1 were reviewed between 1998 and 2018. Pre and post diagnosis clinical picture, biochemical exams, neuroimaging, nutritional status and intake of lysine, proteins, calcium, iron and zinc were recorded. **RESULTS:** 21 AG-1 are in active control (13 men/9 women), diagnostic age: 0.6-48 months of age; 9/21 cases debuted with AEC and 12/21 did not have AEC. Of the cases without AEC: 8/12 have neurological involvement (extrapyramidal symptoms) and 4/12 have a normal development. Prior to diagnosis, 20/21 cases had macrocephaly, delayed psychomotor development and/or abnormal movements. According to nutritional status: 8/21 are eutrophic, 9/21 have malnutrition, 2/21 are overweight and 2/21 are at risk of malnutrition. Eleven have a gastrostomy. The diagnosis was made by MS/MS, confirming with presence of GA, 3OH-GA. Only 1 case was detected by an expanded newborn screening. 8/21 cases GA-1 are <6 years old and are on a lysine restricted diet with a contribution of 0.6±0.3 g intact prot/wt/day, 0.9±0.1 g prot of SF/wt/day and are supplements with: riboflavin, L-carnitine (x 76±39 mg/wt/day) and 12/21 are> 6 years old (6 are on diet and SF and 6 are without SF, but with low lysine diet. **CONCLUSIONS:** GA-1 has a variable clinical presentation, and the neurological compromise can occur despite not having experienced AEC. The GA-1 in active control comply with the contribution of protein, energy, lysine and micronutrients according to the protocol for GA-1 and according to RDA. The GA-1> 6 years (5/12) receive doses of L-carnitine x 29±10mg/wt/day, and maintain free carnitine level above 70%.