# Poor Access to Liver Transplantation and Survival of Children With Acute Liver Failure, Acute-on-chronic Liver Failure or Chronic Liver Disease

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## Abstract

We describe the survival of children with acute liver failure (ALF), chronic liver disease (CLD), or acute-on-chronic liver failure (ACLF) with poor access to liver transplantation (LT). A retrospective cohort study of 42 patients <18 years of age was conducted in the Hospital Civil de Guadalajara "Dr. Juan I. Menchaca". The median age was 76 months; 57.1% were female, 40.5% presented with ALF, 35.7% with CLD, and 23.8% with ACLF. Also, 38.1% (16/42) presented liver disease of unknown etiology. Death occurred in 45.2%; 14.3% were transferred to another hospital, and none received LT. Mortality in ALF, CLD, and ACLF was 76%, 0%, and 60%, respectively. In the survival analysis, within the first 20 months after diagnosis, the mortality rate was greater than 50% with ALF. The importance of having referral programs that perform liver transplantation is highlighted by the poor prognosis of the patients, despite conservative treatment.

**Keywords:**acute; end-stage liver disease; liver diseases; liver failure.

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# Awake prone positioning in acute hypoxaemic respiratory failure: An international expert guidance

## Abstract

**Background:**Awake prone positioning (APP) of non-intubated patients with acute hypoxaemic respiratory failure (AHRF) has been inconsistently adopted into routine care of patients with COVID-19, likely due to apparent conflicting evidence from recent trials. This short guideline aims to provide evidence-based recommendations for the use of APP in various clinical scenarios.

**Methods:**An international multidisciplinary panel, assembled for their expertise and representativeness, and supported by a methodologist, performed a systematic literature search, summarized the available evidence derived from randomized clinical trials, and developed recommendations using GRADE (Grading of Recommendations, Assessment, Development, and Evaluation) methodology.

**Results:**The panel strongly recommends that APP rather than standard supine care be used in patients with COVID-19 receiving advanced respiratory support (high-flow nasal cannula, continuous positive airway pressure or non-invasive ventilation). Due to lack of evidence from randomized controlled trials, the panel provides no recommendation on the use of APP in patients with COVID-19 supported with conventional oxygen therapy, nor in patients with AHRF due to causes other than COVID-19.

**Conclusion:**APP should be routinely implemented in patients with COVID-19 receiving advanced respiratory support.

# [Association of illicit drug use in pregnant women and congenital syphilis in a public hospital in Mexico]

[Article in Spanish]

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## Abstract

We conducted a nested case-control study within a cohort with the aim of studying the association between illicit drug use and congenital syphilis (CS). Cases were diagnosed based on treponemal and non-treponemal tests conducted both in the mother and the newborn (NB). Multivariate analysis with logistic regression was performed. A total of 6171 births with a mean gestational age of 37.8 weeks were recorded and 62 CS events were diagnosed (incidence 10.5 events/1000 NB). Associated maternal factors were illicit drug use (OR 14.08, 95% CI 1.19-166.6), <5 prenatal visits (OR 2.9, 95% CI 1.12-7.53), more than two sexual partners (OR 3.76, 95% CI 1.62-8.71) and professional education level (OR 0.06, 95% CI 0.005-0.85). Among the mothers of the cases presented, the prevalence of illicit drug use was 22.6% and the most frequent drugs were methamphetamines and cannabis.

**Keywords:**Atención prenatal; Congenital syphilis; Drogas ilícitas; Illicit drugs; Prenatal care; Sífilis congénita.

# Systolic blood pressure and the risk of kidney replacement therapy and mortality in patients with chronic kidney disease stage 4-5

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## Abstract

Introduction In patients with chronic kidney disease stage 4 and 5 (CKD stages 4-5) without dialysis and arterial hypertension, it is unknown if the values of systolic blood pressure (SBP) considered in control <120 mmHg are associated with kidney replacement therapy (KRT) and mortality. Methods In this retrospective cohort study, hypertensive CKD stages 4-5 patients attending the Renal Health Clinic at the Hospital Civil de Guadalajara were enrolled. We divided them into those that achieved SBP < 120 mmHg (controlled group) and those who did not (>120 mmHg), the uncontrolled group. Our primary objective was to analyze the association between the controlled group and KRT; the secondary objective was the mortality risk, and if there were subgroups of patients that achieved more benefit. Data were analyzed using Stata software, version. 15.1. Results During 2017 to 2022 a total 275 hypertensive CKD stages 4-5 patients met the inclusion criteria for the analysis, 62 in the controlled group and 213 in the uncontrolled group; mean age 61 years, 49.82% were male, SBP was significantly lower in the controlled group (111 mmHg) compared to the uncontrolled group (140 mmHg), eGFR was similar between groups (20.41 ml/min/1.73m2). There was a tendency to increase the mortality risk in the uncontrolled group (HR 6.47 [0.78-53.27]; p= 0.082) and an association by the Kaplan-Meir analysis (Log-rank p= 0.043). The subgroup analysis for risk of KRT in the controlled group revealed that patients ≥ 61 years had a lower risk of KRT (HR 0.87 [95% CI, 0-76-0.99]; p=0.03, p of interaction = 0.005), but no differences were found in the subgroup analysis for mortality. In a follow-up of 1.34 years, no association was found in the risk of KRT according to the controlled or uncontrolled groups in a multivariate Cox analysis. Conclusion In a retrospective cohort of patients with CKD stages 4-5 and hypertension, SBP >120 mmHg was not associated with risk of KRT but could be associated with the risk of death. Clinical trials are required in this group of patients to demonstrate the impact of reaching the SBP goals recommended by the KDIGO guidelines.

# Pilot study on the efficacy and safety of berberine in people with metabolic syndrome and human immunodeficiency virus infection

## Abstract

**Background:**Antiretroviral therapy has increased the life expectancy of people living with HIV. However, this increase is not free of comorbidities, and metabolic syndrome is one of the most prevalent. Berberine is an alkaloid nutraceutical that has been shown to ameliorate metabolic disorders such as prediabetes, polycystic ovary syndrome, and non-alcoholic fatty liver disease. However, it has not been tested in HIV infection. Therefore, we conducted a randomized controlled trial to evaluate the efficacy of berberine in improving metabolic syndrome.

**Methods and results:**In this double-blind, placebo-controlled trial, adults living with HIV under virological suppression and metabolic syndrome received either berberine 500 mg TID or placebo for 20 weeks. The primary outcomes were a composite of weight reduction, insulin resistance decrease, and lipid profile improvement. A total of 43 participants were randomized (22 in the berberine group and 21 in the placebo group); 36 participants completed the follow-up and were analyzed. The berberine group showed a reduction in weight and body mass index, lower insulin resistance, and a reduction in TNF-alpha. The control group had higher total cholesterol, c-LDL, and IL-6 concentration.

**Conclusion:**In people living with HIV under virological suppression, berberine was safe and improves clinical and biochemical components of metabolic syndrome. However, further studies with more participants and longer intervention periods need to be explored.

**Keywords:**Human immunodeficiency virus infection; berberine; cardiovascular risk; dyslipidemia; insulin resistance; metabolic syndrome.

# MTSS2-related neurodevelopmental disorder: Further delineation of the phenotype

## Abstract

MTSS2-related neurodevelopmental disorder (MTSS2-related NDD) (MIM 620086) is characterized by intellectual developmental disorder with ocular anomalies and distinctive facial features (IDDOF). The only existing report to date described five individuals who exhibited an identical de novo c.2011C>T (p.Arg671Trp) variant in the MTSS2 gene. Herein, we report a new case of MTSS2-related NND in a male dizygotic twin who presented with IDDOF and severe intellectual disability. This patient also displayed additional clinical features, including low functioning autism, hypothyroidism, duodenal obstruction secondary to Ladd's bands, inguinal hernias, cryptorchidism, transient subperiosteal new bone formation, and short stature with delayed bone age, which had not been previously reported in association with the MTSS2-related NDD. Exome sequencing identified the recurrent c.2011C>T (p.Arg671Trp) variant in the MTSS2 gene. The mother and the other twin tested negative for the pathogenic variant, while the father's participation in the study was unavailable. This case confirms that the MTSS2-related NDD is caused by the recurrent MTSS2 missense variant p.Arg671Trp. The novel findings identified in our patient expand the phenotypic spectrum associated with this new autosomal dominant entity, but further studies on its genetic and clinical manifestations are still needed.

**Keywords:**ABA1; Intellectual developmental disorder; MTSS1L; MTSS2; Ptosis.

# Nanocatalytic performance of pectinase immobilized over in situ prepared magnetic nanoparticles

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## Abstract

Immobilization of enzymes is one of the protein engineering methods used to improve their thermal and long-term stabilities. Immobilized pectinase has become an essential biocatalyst for optimization in the food processing industry. Herein, nanostructured magnetic nanoparticles were prepared in situ for use as supports to immobilize pectinase. The structural, morphological, optical and magnetic features and the chemical compositions of the nanoparticles were characterized. Nanoparticle agglomeration and low porosity were observed due to the synthetic conditions. These nanoparticles exhibited superparamagnetic behavior, which is desirable for biotechnological applications. The maximum retention rate for the enzyme was observed at pH 4.5 with a value of 1179.3 U/mgNP (units per milligram of nanoparticle), which was equivalent to a 65.6% efficiency. The free and immobilized pectinase were affected by the pH and temperature. The long-term instability caused 40% and 32% decreases in the specific activities of the free and immobilized pectinase, respectively. The effects of immobilization were analyzed with kinetic and thermodynamic studies. These results indicated a significant affinity for the substrate, a decreased reaction rate, and improved thermal stability of the immobilized pectinase. The reusability of the immobilized pectinase was preserved effectively during cycling, with only a 21.2% decrease in activity observed from the first to the last use. Therefore, alternative magnetic nanoparticles are presented for immobilizing and maintaining the thermostability of pectinase.

**Keywords:**Cross-linking; Enzyme immobilization; Magnetic nanoparticles; Pectinase.

# Methemoglobinemia in Hemodialysis Patients due to Acute Chlorine Intoxication: A Case Series Calling Attention on an Old Problem

## Abstract

**Introduction:**Hemodialysis uses municipal water that must be strictly purified and sterilized to be used for that procedure. Large amounts of decontaminants are often used, such as chlorine, and if these compounds are not subsequently removed they can be transferred to the blood of patients causing complications including methemoglobinemia.

**Methods:**In this case series study, dialysis patients in one unit were evaluated. We reviewed clinical characteristics and laboratory findings obtained on the day when the water supply was disinfected with chlorine, with the aim to quantify methemoglobin concentrations. Our objective was to characterize the clinical presentation and management of patients who presented with methemoglobinemia on a specific index day. We also reviewed reported cases in the literature regarding this underreported complication.

**Results:**Eight patients who presented with chlorine intoxication were evaluated. The methemoglobin concentrations were between 1.3% and 7.9% (reference value 0-1%). We believe this to be caused by water containing 0.78 mg/L of total chlorine. Seven patients presented with cyanosis, 4 with dizziness, 6 with dark brown blood, 4 with dyspnea, and 4 with headache and hemolytic anemia. Subjects were treated with supplemental oxygen, methylene blue, intravenous vitamin C, blood transfusions, and increased doses of erythropoietin. No patient died, and all continued with their usual hemodialysis sessions.

**Conclusion:**Acute chlorine intoxication transferred by the water used during hemodialysis sessions can present with methemoglobinemia accompanied by cyanosis, oxygen desaturation, and hemolytic anemia. Chlorine levels should be carefully monitored in the water used for hemodialysis treatment.

**Keywords:**Chlorine; Hemodialysis; Methemoglobinemia.

# Ataxia due to a COQ8A Novel Variant in Primary Coenzyme Q10 Deficiency

No abstract available

**Keywords:**COQ8A; ataxia; coenzyme Q10; movement disorders.