Child classes as regards spirometry pattern, where 40% of Child C patients showed mild restrictive pattern. There was highly significant difference between positive HPS and negative HPS patients as regards PaO2, A-aDO2, VC (L), FVC (L), FEV-1(L/sec), and FEF25-75% (L/sec). There was highly significant positive correlation between A-aDO2 and each of age, INR and portal vein diameter. There was highly significant negative correlation between A-aDO2 and each of serum albumin, PaO2, VC, FVC, FEV-1 and FEF 25-75%. A cutoff level ≥ 18.4 mmHg for A-aDO2 could detect positive HPS with a sensitivity of 100%, specificity of 96.3%, PPV of 97.1% and NPV of 100%.

Conclusion: A cutoff level ≥ 18.4 mmHg for A-aDO2 could detect HPS. There is significant correlation between the severity of chronic liver disease and the presence of HPS.

The frequency, clinical patterns, nutritional status and health related quality of life in adults with Gilbert’s syndrome: a prospective longitudinal study

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Goals: Investigation of the frequency clinical spectrum, genetic profile and health related quality of life (HRQOL) in Egyptian with Gilbert syndrome (GS).

Background: GS is an inherited disorder of bilirubin glucuronidation characterized by unconjugated hyperbilirubinaemia, GS has not been previously investigated in Egyptians.

Study: An initial cross-sectional study was conducted to assess the frequency of Gilbert syndrome among Egyptians. Patients fulfilling the criteria of GS were longitudinally followed and further investigated for the risk factors for hyperbilirubinemia, health related quality of life (HRQOL) and UGT1A1 polymorphisms.

Results: Of screened patients, 83 individuals fulfilled the criteria of GS. Jaundice was the only manifestation in 47/83 (56.627%) subjects with GS jaundice was associated with fatigue, abdominal pain and dyspepsia 43.37%, 22.2% and 25.3% of patients respectively. The risk factors for hyperbilirubinemia included continuous fasting > 10 hours, infections, extensive physical exercise, surgical interventions, psychologic stress and menstruation, pregnancy and oral contraceptives in women. Lipid profile abnormalities were detected in 36 (43.37%) of subjects. The SF-36v2 and CLDQ scores were low during the jaundice attacks in 69 (83.132%) subjects with GS. UGT1A1*28 polymorphisms were detected in 51 (61.45%), 39 (41.94%) and 19 (19%) individuals with GS, family members of GS patients and healthy individuals respectively.

Conclusion: Gilbert syndrome is an underestimated cause of fluctuating jaundice in Egyptians. Many of the identified risk factors for hyperbilirubinemia maybe avoided. Several domains of health related quality of life are affected in patients with Gilbert syndrome. UGT1A1*28 polymorphisms may be useful in identification of asymptomatic GS cases.