Perioperative anesthetic management of patients with Gaucher disease: a 10-year retrospective survey

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Background and Goal of Study: Gaucher disease, a rare lysosomal storage disorder caused by an enzyme deficiency, results in hepatosplenomegaly and consequent cytopenia as well as lung and skeletal pathology in some cases and more rarely, neurological involvement. Enzyme replacement therapy is standard management for visceral signs in symptomatic patients.

Methods and results: In the past decade (retrospective chart review) 86 surgeries in 59 patients were performed at our clinic, of which two patients had the neuropathic form, not carrying a N370S mutation on at least one allele. Mean Severity Score Index was 11.7 points (range 0-27 when 30 is the worst possible score) indicative of moderate involvement. One third of the patients underwent orthopedic surgeries and one-eighth obstetric.

Discussion: In addition there were 20 invasive procedures (colonoscopies and gastroscopies) that did not require anesthesia. 50% of the surgeries employed general anesthesia with no complications noted. Similarly, various surgeries employing regional anesthesia (spinal, epidural and peripheral nerve block) were uneventful. Because of concurrent anemia (12%) or thrombocytopenia (20%), 26% of patients received blood products during or after the surgery. Most cases only required standard monitoring but in 20%, monitoring including inserting a peripheral arterial line for direct pressure measurements and blood sampling. Thromboelastography (TEG) is an important part of the perioperative management of these patients secondary to potential hypocoagulation state.

Conclusion : The good results we have seen are in great measure because of improvement in hematological parameters because of enzyme therapy. In patient with platelets $>50,000/\text{mm}^3$ and normal TEG regional anesthesia is feasible. No airway problems as a result of Gaucher diseases were recognizing.

References:

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