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241 AQX-1125, a Modulator of the SHIP1/PI3K Pathway, Suppresses Chemotaxis and Inflammation

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RATIONALE: AQX-1125, a novel small-molecule allosteric activator of SHIP1, was evaluated for *in vitro* and *in vivo* anti-inflammatory activity. **METHODS:** AQX-1125 was tested in an *in vitro* Boyden chamber chemotaxis assay. The efficacy of AQX-1125 on reducing ear thickness and myeloperoxidase content was evaluated *in vivo* in the phorbol 12-myristate 13-acetate (PMA)-induced ear edema mouse model. AQX-1125 was administered orally for 3 days at 1, 10 and 30 mg/kg prior to application of PMA. The efficacy of AQX-1125 was also tested in a rat ovalbumin-mediated airway inflammation model, administered orally for 4 days at 0.1, 1 and 10 mg/kg before airway challenge. The degree of inhibition of leukocyte infiltration and mediator release in the bronchiolar lavage fluid (BALF) was measured.

RESULTS: AQX-1125 is a potent inhibitor of *in vitro* leukocyte chemotaxis. *In vivo*, AQX-1125 at 30 mg/kg significantly reduced ear edema and myeloperoxidase content in the PMA model. At 1 mg/kg, AQX-1125 significantly reduced the total number of leukocytes recovered in the BALF of animals sensitized and challenged with ovalbumin. AQX-1125 also reduced the level of the inflammatory mediators IL-1 α and IL-11, in the BALF.

CONCLUSIONS: AQX-1125 potently inhibits leukocyte chemotaxis *in vitro* and myeloperoxidase (neutrophil) accumulation *in vivo*. AQX-1125 also inhibits leukocyte accumulation and inflammatory mediator release in the BALF in a model of allergic airway inflammation. These data suggest that AQX-1125 has clinical potential for treatment of allergic and inflammatory diseases such as asthma and COPD.

242 Lipid Profile in Patients With Primary Immunodeficiency

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RATIONALE: Chronic diseases can present higher inflammatory and oxidative stress leading to a greater risk of development abnormalities in lipid metabolism.

METHODS: In a cross-sectional study, we evaluated the lipid profile of 48 patients with PID who have been treated with regular intravenous immunoglobulin, at the outpatient clinic at UNIFESP/EPM. PID diagnosis was based on PAGID/ESID criteria. The nutritional status was determined by body mass index (BMI) and height for age index (H/A) according to WHO.

RESULTS: We evaluated 48 PID patients 56.3% (27/48) of them children and adolescents with a mean age of 10.7 \pm 4.6 years in this group, and 63.0% (17/27) of whom were males. The adults mean age \pm SD was 32.87 \pm 9.73 years, with 52.4% male. Humoral ID was the predominant PID with 68.8% (33/48) followed by 16.7%(8/48) cellular immunodeficiency and 14.6% had Ataxia-Telangiectasia. Regarding children and adolescents nutritional status, 37.0%(10/27) were of short stature and 25.9% (7/27) were overweight. In adults, 23.8%(5/21) had malnutrition and were overweight. According to the lipid profile, 18.5%(5/27) of children and adolescents had high levels of total cholesterol and LDL-cholesterol, and 51.9%(14/27) had low HDL-cholesterol. For adults, we observed abnormalities in 33.3%(7/21) who presented high levels of total cholesterol, 19.0%(4/21) for triglycerides, 9.5%(2/21) for LDL-cholesterol and 61.9% (13/21) had low levels of HDL-cholesterol.

CONCLUSION: Nutritional status, high cholesterol levels with low levels of HDL-cholesterol showed abnormalities in a high percentage of PID patients

243 Improving Adherence with Initiating New Prescriptions for Inhaled Corticosteroid and EpiPen

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RATIONALE: A quality improvement project to improve medication adherence by assessing barriers to obtaining and administering new prescriptions for inhaled corticosteroid and EpiPen after RN education in the pediatric pulmonary/allergy office.

METHODS: After education in the office, RN placed a one week follow-up call to the home addressing adherence and barriers to adherence with the following questions:

1. Have you filled the prescription for the medication?
2. Is your child taking the medication? (Does not apply to EpiPen)
3. What dose is your child taking?
4. Are there any barriers to taking the medication?
5. Do you have any concerns or questions about the medication?

RESULTS: Adherence as reported by parents ranged from 75% to 85% over the course of the project. This exceeded the 50% predicted based on a literature search. The project impacted care by increasing the nursing staff's awareness of barriers to adherence, allowing staff to be proactive in addressing those issues.

CONCLUSIONS: This quality improvement project can be interpreted as a successful initiative.

244 Addressing the Need for Improved Patient and Nurse Education on Subcutaneous Immunoglobulin: Enhancing Patient Experiences and Nurse Support

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RATIONALE: Nursing school curriculums currently do not provide training for subcutaneous immunoglobulin (SCIG) infusion therapy, and educational materials by SCIG manufacturers for nurses and patients with primary immunodeficiency (PI) are not sufficient. This has led to suboptimal patient experiences, which may contribute to reduced adherence to therapy and increased side effects. To address this unmet need, a panel of nurses was convened to develop a comprehensive self-administration educational tool for both patients and nurses who are unfamiliar with SCIG therapy.

METHODS: A group of 9 specialty infusion nurses from both academic and homecare settings participated in an advisory board meeting. Current challenges in patient compliance and best practices in patient and nurse education were discussed. An adult educator advised on various patient learning styles.

RESULTS: A modular learning tool was developed and reviewed by the panel. Separate tools were developed and tailored for pediatric, adult and elderly patient subgroups. A group of patients with PI validated the final educational packages. A separate modular resource was developed to support nurse training and education on SCIG treatment.

CONCLUSIONS: Robust educational programs on SCIG therapy are needed to enhance patient experiences and outcomes and improve nurse support. Effective patient and nurse training tools will likely lead to improved adherence to therapy and help patients reach infusion independence.