To date, BION-1301 has been well-tolerated in IgAN patients (n=10). BION-1301 treatment results in clinically meaningful proteinuria reductions within 3 months in patients across a wide range of disease severities. Median baseline 24-h urine protein excretion*: 1.22 g/day (range: 0.74 - 6.47 g/day). BION-1301 durably reduces IgA, IgM, and to a lesser extent, IgG in patients with IgAN.

No anti-drug antibodies observed in patients with IgAN to date. Rapid and durable reductions in free APRIL confirm effective target neutralization sustained through 1 year. BION-1301 pharmacokinetics in patients with IgAN is consistent with previous experience in healthy volunteers. BION-1301 produces sustained reductions in serum levels of fAPRIL, IgA, and IgM, and was measured by immunoturbidimetry. UPCR was assessed from 24-hour urine collections.

The Phase 1 study (NCT03043318) comprises 3 parts: Parts 1 and 2 are single- and multiple-dose ascending studies of BION-1301 in IgAN. Part 3 is an ongoing, open-label design in approximately 40 IgAN patients. Cohort 1 enrolled 10 patients to receive BION-1301 at 450mg once every 2 weeks intravenously. Patients in Cohort 3 will transition from Part 2 to receive BION-1301 600mg every 4 weeks after completing 12 weeks of therapy in Cohort 2. Cohort 2 is currently enrolling patients to receive BION-1301 at 600mg once every 2 weeks for up to 52 weeks via subcutaneous administration.

**Mechanism of APRIL and BION-1301 in IgAN Nephropathy**

- BION-1301 is a novel humanized monoclonal antibody (mAb) against APRIL. No anti-drug antibodies observed in patients with IgAN.
- Durable reductions in free APRIL confirm effective target neutralization sustained through 1 year.
- BION-1301 pharmacokinetics in patients with IgAN is consistent with previous experience in healthy volunteers.
- BION-1301 produces sustained reductions in serum levels of fAPRIL, IgA, and IgM, and was measured by immunoturbidimetry. UPCR was assessed from 24-hour urine collections.

**Methods**

The Phase 1 study (NCT03043318) comprises 3 parts: Parts 1 and 2 are single- and multiple-dose ascending studies of BION-1301 in IgAN. Part 3 is an ongoing, open-label design in approximately 40 IgAN patients. Cohort 1 enrolled 10 patients to receive BION-1301 at 450mg once every 2 weeks for up to 52 weeks intravenously. Patients in Cohort 3 will transition from Part 2 to receive BION-1301 600mg every 4 weeks after completing 12 weeks of therapy in Cohort 2. Cohort 2 is currently enrolling patients to receive BION-1301 at 600mg once every 2 weeks for up to 52 weeks via subcutaneous administration.

**Results**

BION-1301 has been well-tolerated in IgAN patients receiving 450mg IV dose every 2 weeks for 12 weeks with no SAEs or treatment-related AEs. Consistent with PD responses previously reported in HN, durable reductions in serum levels of NFκB and immunoglobulins were observed in IgAN patients. Clinically meaningful reductions in proteinuria were seen as early as 12 weeks and were associated with the reduction in Gd-IgA1 levels.

**Disclosures for Presenting Author**


**References**


