Results: The interim analysis includes 52 CAPS patients with prior long-term CAN treatment (43.1% females) enrolled by September 2018. 44.2% of the patients participated in the [i-Confident study previously. The mean age was 20.7 years (4.0–79.0 years) at baseline and the mean duration of prior CAN treatment was 5.4 years (0.0–11.0 years). 40 patients (76.9%) were diagnosed with MWS, the other patients had FCAS (2%), NOMID/CINCA (7) or atypical CAPS (1) (subtype diagnosis of 2 patients not available). Mutations in Nod-like receptor family pyrin domain-containing-3 were identified in 34 patients (68.0%) including E311K (7), Q703K (5), V198M (5), T348M (5), A439V (4). A baseline screening revealed sensorineural hearing loss (82.0%), papillitis (94.0%) and neurological symptoms (71.4%) in patients which had not been detected previously. The majority of patients had no disease related symptoms at baseline and 6 months demonstrating sustained remission in patients receiving long-term CAN treatment. The following disease related symptoms (mild/moderate and severe) were observed in the analysis cohort (N=31) at baseline and 6 months, respectively – disease symptom: baseline (mild/moderate, severe) vs. 6 months (mild/moderate, severe) – uveitis: 19.4%, 6.5% vs. 25.8%, 0.0%, arthritis: 32.3%, 0.0% vs. 29.0%, 9.7%, myalgia: 9.7%, 0.0% vs. 16.1%, 0.0%, headaches: 22.6%, 9.7% vs. 19.4%, 19.4%, conjunctivitis: 32.3%, 3.2% vs. 12.9%, 6.5%, abdominal pain: 9.7%, 6.5% vs. 22.6%, 9.7%. Patients’ assessment of disease activity and fatigue did not change between baseline and 6 months. However, at baseline 45.5% and after 6 months 76.0% of patients had no impairment of social life by the disease. Serious adverse events were reported for 2 patients (tornillas, delivery at week 31).

Conclusion: The RELIANCE study longitudinally monitors the stability of effectiveness of CAN in patients with monogenic periodic fever syndromes. An initial interim analysis including the CAPS subgroup which had prior CAN treatment showed that CAN is an effective and safe treatment in those patients.

Disclosure of Interests: · J. B. Kuenemmerle-Deschner Grant/research support from: Jasmin Kuenemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis, Consultant for: Jasmin Kuenemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis, Speakers bureau: Jasmin Kuenemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis. Speakers bureau: Jasmin Kuenemmerle-Deschner is an employee of University of Tuebingen, Germany, and received consultants/speakers fees from Novartis and SOBI pharmaceuticals and grant support from SOBI and Novartis.

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Disclosure of Interests: None declared

Figure 1. Treatment Assignments and Withdrawal in the Intention-to-Treat Population.