

	Exclusion criteria*
1	Treatment with nitisinone within 3 months of randomization
2	Participation in another clinical study within 3 months of randomization
3	Known allergy to nitisinone or any of the constituents of the investigational product
4	Female patient of child-bearing potential not using a reliable method of contraception
5	Currently pregnant or lactating
6	Current malignancy
7	Uncontrolled hypertension (blood pressure greater than 180 mmHg systolic or greater than 95 mmHg diastolic)
8	Unstable cardiovascular disease
9	Serum potassium < 3.0 mmol/L
10	eGFR < 60 mL/min/1.73 m ²
11	ALT > 3 x upper limit of normal
12	Haemoglobin < 10.0 g/dL
13	Platelets < 100 x 10 ⁹ /L
14	Total white blood count < 3.0 x 10 ⁹ /L or neutrophil count < 1.5 x 10 ⁹ /L
15	History of alcohol or drug abuse
16	Psychiatric or somatic illness that interferes with compliance or communication with health care personnel
17	Foreseeable inability to cooperate with given instructions or study procedures
18	Any other medical condition which in the opinion of the investigator makes the patient unsuitable for inclusion

Supplementary Table S2. SONIA 2 exclusion criteria. *The presence of any of the following excluded a patient from inclusion in the study.