REgistry of Pediatric Sjögren syndrome in Umcg- LongiTudinal (REPSULT)

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Ethical review	Approved WMO
Status	Recruiting
Health condition type	Autoimmune disorders
Study type	Observational non invasive

Summary

ID

NL-OMON49266

Source
ToetsingOnline

Brief title
REPSULT

Condition

- Autoimmune disorders

Synonym
Sjogren syndrome

Research involving
Human

Sponsors and support

Primary sponsor: Universitair Medisch Centrum Groningen
Source(s) of monetary or material Support: reumanl

Intervention

Keyword: Follow up, Pathofysiology, Pediatric, Sjögren syndrom

Outcome measures

Primary outcome

Aims:

1. Define disease presentation of children with SS-like complaints

2. Evaluate current adult classification criteria in pedSS patients

3. Analyze the genetic constitution of pedSS

4. Assess essential biomarkers (of adult SS) in children with SS

5. Analyze and describe the clinical course and HRQoL of pedSS

6. Identify predictors of MALT lymphoma in pedSS

Parameters

Clinical and patient-reported parameters, salivary and tear gland function,

ultrasound and histopathology of salivary glands, laboratory parameters,

immunological parameters in serum, saliva and tears, and genetic markers.

Secondary outcome

na

Study description

Background summary

Sjögren syndrome (SS) is a lifelong, systemic autoimmune disease that has major impact on daily activities and quality of life. In adult SS the salivary and lacrimal glands are most commonly affected, resulting in xerostomia (dry mouth) and keratoconjunctivitis sicca (dry eyes). Pediatric-onset Sjögren syndrome

(pedSS) more often presents with other symptoms such as recurrent parotitis and renal and neurological impairment. The underlying pathophysiological mechanism in pedSS may be different from adult SS, with a more prominent role for genetic diseases. The consequences of pedSS on daily life has not been studied. For this reasons there is a urgent need to evaluate the value of the current classification criteria in pedSS and further research in the pathophysiological mechanisms and genetic background.

Study objective

Our hypothesis is that pedSS has a different presentation and pathophysiology than adult SS, with more genetic involvement. To test this hypothesis, we define the following specific aims: define disease presentation of children with SS-like complaints, analyze and describe the clinical course and HRQoL of pedSS, evaluate current adult classification criteria in pedSS patients, assess essential biomarkers (of adult SS) in children with SS, analyze the genetic constitution of pedSS and identify predictors of MALT lymphoma in pedSS.

Study design

The design will be a longitudinal observational prospective cohort study. All consecutive patients with confirmed or probable SS before the age of 17 years, who visit the (outpatient) clinic of the department of Pediatric Rheumatology in the UMCG are considered for this study. Inclusion will take place during 10 years and all patients will be followed up for 10 years. Visits according to a fixed protocol will be performed at baseline and every year thereafter, with more extensive evaluation after 1, 2, 5 and 10 years of follow up (Figure 1). If indicated, additional visits may be scheduled as part of standard care, i.e. because of high disease activity or treatment with systemic immunosuppressant drugs. This additional visits can also take place in the referring hospital in a shared care construction. For patients with probable pedSS, the clinical suspicion of pedSS might be lowered during follow-up, making yearly visits unnecessary. From the moment yearly visits will be stopped and these patients will be followed up only in year 1, 2, 5 and 10. When patients reach the age of 18 the clinical care will be transferred to the adult rheumatologist, but they will continue to participate in the REPSULT cohort.

Study burden and risks

Participants will visit the outpatient clinic in the context of clinical care, and will not bring additional visits to the hospital. Extra measurements (which do not concern the regular clinical follow-up of pedSS patients) are questionnaires on xerostomia, health related quality of life and school and collection of extra blood (during routine venepuncture), saliva and tears and a yearly ultrasound of the parotic glands (non-invasive procedures). Diagnostic procedures or treatment will not be postponed. Overall, the burden for the

participants is low and the risk of participation is negligible as no extra invasive procedures will be done. Patients will not experience any direct benefits from participation in this cohort. However, the results of this cohort might improve the quality of care for patients with (pediatric) SS in the future. Therefore, we assume that the advantages of participation in this cohort outweigh the burden of participation.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adolescents (12-15 years)
Adolescents (16-17 years)
Adults (18-64 years)
Children (2-11 years)
Elderly (65 years and older)

Inclusion criteria

Confirmed or probable SS diagnosed before the age of 17 years.

Exclusion criteria

Subjects who are impaired or incapable of completing cohort-related assessments
Serious comorbidity or laboratory abnormalities that, in the opinion of the investigator, unacceptably increase the burden or risk of participation in the cohort.

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)
Control:	Active
Primary purpose:	Basic science

Recruitment

NL

Recruitment status:	Recruiting
Start date (anticipated):	04-06-2020
Enrollment:	50
Type:	Actual

Ethics review

Approved WMO

Date:	29-04-2020
Application type:	First submission
Review commission:	METC Universitair Medisch Centrum Groningen (Groningen)

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL71206.042.19