Abstract 1244 Table 1 Comparison of average HbA1c (mmol/mol) pre- and post-lockdown grouped by baseline HbA1c

Group	Number of Subjects	Pre-LD (1)	Post-LD (2)	Av-Post-LD (3)	Improved 1-2	Improved 1-3	Improved 2-3
>69	38	86.67	76.94	76.49	Yes	Yes	Yes
64-69	25	65.69	61.82	61.07	Yes	Yes	Yes
58-63	44	60.14	58.31	59.78	Yes	Yes	No
53-57	23	55.35	54.23	54.63	Yes	Yes	No
48-52	18	50.51	52.25	52.59	No	No	No
<48	13	42.07	44.24	46.50	No	No	No

highlighting the pandemic's disruption to the care of patients with long-term conditions. Contrary to guidance, 14.73% of patients were not tested in the nine months post-lockdown, potentially reflecting reluctance to attend hospital during the pandemic. Nevertheless, where results were available, lockdown seemed to have a positive impact on HbA1c despite the enforced use of telemedicine, unless the patient was already close to target. It is possible that lockdown provided more routine and parental supervision for those with initially high HbA1c but disrupted the successful routine and sporting activities of those initially closer to target.

British Paediatric Respiratory Society

1245

IMMUNOLOGICAL FINDINGS AND RESPIRATORY MANIFESTATIONS IN ATAXIA-TELANGEICTASIA: A SCOPING REVIEW

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Background Ataxia-telangiectasia (A-T) is a rare autosomal recessive, multi-system, neurodegenerative disease due to mutations in the ataxia-telangiectasia mutated (ATM) gene. It causes neurological impairments, immunodeficiency leading to serious recurrent infections, and malignancy.

There are two types of A-T; the more severe classical A-T that presents in early childhood, and the less severe variant A-T that presents later in childhood, or adulthood.

People with classical A-T generally present at approximately 18 months of age with a cerebellar gait ataxia, are wheelchair-bound by 10 years and rarely live beyond their twenties, with death mainly due to malignancy or lung disease.

People with variant A-T present later in childhood, or as an adult, and have a milder phenotype with a longer survival. **Objectives** To document the immunological and respiratory manifestations of A-T.

Methods 17 searches were carried out in each of 5 databases (Ovid SP (Medline), EMBASE, Web of Science, PubMed, Scopus). The Cochrane Library was also searched. The search protocol is available.

The inclusion criteria were all dates, languages and ages; humans; and clinical relevance. The exclusion criteria were: no reference to A-T, not an original article, animal studies, not clinically relevant.

Results Search returned 194,890 articles; 14,622 titles and abstracts were reviewed after removing 180,268 duplicates.

Full text review of 1,163 articles was performed and 1,039 studies were included (13,459 exclusions, 124 excluded after full text review).

The most frequently reported immunoglobulin abnormality was low IgA in the classical group, followed by high IgM. The majority of immunoglobulin results reported in the variant group were normal. The most likely abnormality in the variant group was high IgM. Very limited age data were reported.

Other common immunoglobulin abnormalities in the classical group included low IgG2 (n=283), low IgG3 (n=44) and low IgG4 (n=86).

699 cases in the classical group received replacement immunoglobulin, starting at a median age (n=40) of 4 years 6 months (range 2 months to 16 years 0 months, IQR 3 years 0 months to 8 years 0 months). No cases in the variant group were reported to be on replacement immunoglobulin. 230 cases in the classical group and 3 in the variant group were reported to have received prophylactic antibiotics.

Recurrent infections were reported in 1118 cases in the classical group and 18 in the variant group. The most frequently reported recurrent infection was sinopulmonary infection in the classical group (n=570) and lower respiratory tract infection/pneumonia in the variant group (n=11).

Non-infectious respiratory manifestations reported in the classical group included bronchiectasis (n=228), interstitial lung disease (n=35), unspecified chronic lung disease (n=3), pulmonary fibrosis (n=7), and pneumothorax (n=50). Age data (where available) will be reported on these manifestations. No non-infectious respiratory manifestations were reported in the variant group.

Conclusions There is a wide variety of infectious and non-infectious respiratory manifestations, and immunological abnormalities reported in classical A-T. There are fewer immunologic and respiratory manifestations reported in variant A-T.

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British Paediatric and Adolescent Bone Group

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JOINT EFFUSIONS: AN INDICATOR OF UNFAVOURABLE OUTCOME OF CRMO IN CHILDREN

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Background Chronic Recurrent Multifocal Osteomyelitis (CRMO) is an autoinflammatory, chronic disease that usually appears in childhood. It can be unifocal or multifocal, and main symptoms are pain and swelling on the area of the affected bones. Most of the times, it has a benign and self-limiting course; however, there is a minority of patients for whom the disease remits often and the symptoms are not well