Abstracts

A MALIGNANCY IN ATAXIA-TELANGIECTASIA: 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 malignancy diagnoses in 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, all languages, all ages, human subjects, and clinical relevance. The exclusion criteria were: no reference to A-T within the article, not an original article, animal studies, article 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).

1826 malignancies were reported in 1643 cases. The most common malignancy in the classical group was non-Hodgkin’s lymphoma (421 cases) and presented at a median age (n=72) of 9 years 8 months (range 6 months to 35 years 6 months, IQR 6 years 0 months to 14 years 0 months). The most common malignancy reported was leukaemia (n=284) presenting at a median age (n=89) of 11 years 0 months (range 1 month to 51 years 0 months, IQR 5 years 6 months to 19 years 0 months), followed by Hodgkin’s disease (n=171).

The most common malignancy in the variant group was breast cancer (n=13) presenting at a median age (n=8) of 34 years 0 months (range 28 years 0 months to 44 years 0 months, IQR 12 years 0 months to 39 years 0 months), followed by leukaemia (n=10) at a median age (n=6) of 9 years 6 months (range 6 years 0 months to 46 years 0 months, IQR 8 years 3 months to 16 years 0 months).

Other results will be presented including the presenting signs and symptoms of leukaemia, non-Hodgkin’s lymphoma and Hodgkin’s lymphoma.

Conclusions A large variety of malignancies are diagnosed in people with A-T at a wide range of ages. Solid tumours are more common in variant A-T and haematological malignancies in classical A-T.

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Children’s Cancer and Leukaemia Group

AN EVALUATION OF WORKING FROM HOME PRACTICES AND EXPERIENCES ON A TERTIARY NICU DURING THE COVID-19 PANDEMIC

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Background During the Covid-19 pandemic, remote ‘working from home’ (WFH) practices were developed on a tertiary neonatal unit to ensure compliance with Covid-19 recommendations around shielding, self-isolating and social distancing, and mitigate anticipated junior doctor staff shortages.

Objectives

- Establish a remote working service for junior doctors unable to attend in person due to Covid-19 restrictions then develop a remote working rota and guideline.
- Enable remote staff to contribute to clinical and non-clinical tasks securely.

Quality Improvement and Patient Safety

To document malignancy diagnoses in A-T.