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Genetic Tumour Syndromes (5th ed.)

1. Forewords and introductions

- Website beta version
- Foreword
- WHO Classification of Tumours: Editorial Board
- Introduction
- Guidelines for the reporting of sequence variants in tumours
- Introduction to Haematolymphoid Genetic Disorders

2. Growth factor receptors and related signalling pathways

3. Oxidative stress response and metabolism

4. Cell cycle and apoptosis pathways

5. DNA repair and genomic stability

6. Telomere maintenance

7. Epigenetic drivers and chromatin remodelling

8. DNA methylation

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WHO Classification of Tumours online

Genetic Tumour Syndromes (5th ed.) // Forewords and introductions

// Introduction to Haematolymphoid Genetic Disorders

Introduction to Haematolymphoid Genetic Disorders

Genetic predisposition is assumed for a variety of haematolymphoid neoplasms, a subset of which is part of the spectrum of several monogenic tumour predisposition syndromes discussed in the present publication. Such monogenic predisposition can occur with or without syndromic features like malformations, dysmorphisms or developmental delay. Moreover, tumour predisposition can be restricted to the haematolymphoid system or affect other organ systems. Correspondingly, the presentation of these monogenic germline predispositions with haematolymphoid neoplasms follows the overall structure of the present issue. Examples include the RAS-MAPK pathway (e.g. Noonan syndrome), MITR pathway (e.g. APDS), transcription factors (e.g. CEBPA, RUNX1, ETV6, GATA2, PRX5, IKZF1 alterations), cell cycle and apoptosis pathways (e.g. Li-Fraumeni syndrome, ALPS), DNA repair and genomic instability pathways (e.g. CMMRD, MBD4, Fanconi anaemia, DDX41, ATM, NBN, BLM), and disorders of telomere maintenance or chromosomal aneuploidy syndromes (e.g. Down syndrome).

The applied pathway-based hierarchical classification leaves few emerging or rare germline tumour syndromes with predisposition to haematolymphoid neoplasms currently unmapped. One example is the SAMD9-related haematologic tumour predisposition syndrome (MIRAGE; MIM 617053). There is evidence that proteins encoded by SAMD9 and its paralog SAMD9L (SAMD9-like) are involved in endosome fusion and that SAMD9 has a role in growth factor signal transduction, but data are not yet sufficient to classify it in the present scheme (Z7182967).

Similarly, the broad field of Inborn Errors of Immunity (IEI) related to predisposition to haematolymphoid neoplasms could not be covered fully here. IEI can contribute to lymphoma predisposition via "direct" tumorigenic mechanisms (e.g. genomic instability in case of ATM mutation) or "indirect" mechanisms such as deficient immune surveillance

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