and atypical MR and to correlate the phenotype and genotype.

METHODS: Exons 3 and 4 of the *MECP2* gene were analyzed by using denaturing gradient gel electrophoresis, sequencing, and gap polymerase chain reaction for (1) 124 children with FXS-like symptoms (102 boys, 22 girls) and 41 children with AS-like symptoms (14 boys, 27 girls) who tested negative for gene variation at the FXS and AS loci, respectively, (2) 23 girls with classical RS and 25 girls with atypical RS, and (3) 11 boys who were referred with possible RS. Statistical analysis (*t* and nonparametrical tests) included correlation of RS clinical severity score (Kerr, 2001) with *MECP2* mutations and frequency of *MECP2* mutations in the various patient categories.

RESULTS: Mutations were detected in 78.3% of classical and 20% of atypical RS cases, respectively. One boy carried the p.R106W mutation, and another boy showed a large rearrangement that required further characterization. Among AS- and FXS-like cases, 7.3% and 2.4% had *MECP2* mutations, respectively, including an X-linked MR case.

CONCLUSIONS: *MECP2* gene analysis provides an appropriate diagnostic tool for RS and contributes additional information for research into MR.

Hematology and Oncology

ASSESSMENT OF BONE MINERAL DENSITY AND MARKERS OF BONE TURNOVER IN CHILDREN UNDERGOING LONG-TERM ORAL ANTICOAGULANT THERAPY

Submitted by Maria Avgeri

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INTRODUCTION: Oral anticoagulants antagonize vitamin K action and potentially impair the carboxylation of osteocalcin, a protein that is essential for normal bone matrix formation.

OBJECTIVE: Our aim was to evaluate bone mineral density (BMD) and bone-turnover markers in 23 children who were undergoing long-term oral anticoagulant therapy (median age: 4 years) and 25 age- and gender-matched controls.

METHODS: BMD (characterized as a *z* score) of the lumbar spine was assessed by using dual energy radiograph absorptiometry. Osteoblast (bone alkaline phosphatase, osteocalcin, and amino-terminal procollagen 1 extension peptide) and osteoclast (urinary calcium and deoxypyridinoline and serum cross-linked C telopeptide) activity markers were measured. Vitamin D (25-hydroxy vitamin D, parathyroid hormone, whole and ionized calcium, phosphorus, and magnesium) and vitamin K (factors II, VII, IX, and X, protein C, protein S, and undercarboxylated osteocalcin [Glu-Oc]) statuses were determined.

RESULTS: Patients presented with higher levels of Glu-Oc, parathyroid hormone, and bone-resorption markers and lower levels of bone-formation markers and 25-hydroxy vitamin D; 52% of them showed signs of osteopenia ($-1.0 >$ BMD *z* score > -2.5). Statistical analysis demonstrated that anticoagulant therapy was an independent predictor of alterations in Glu-OC, osteocalcin, bone alkaline phosphatase, amino-terminal procollagen 1 extension peptide, and serum cross-linked C telopeptide levels.

CONCLUSIONS: Long-term use of coumarin derivatives may cause osteopenia in children with the risk of developing osteoporosis later in life.

IN VITRO ASSESSMENT OF MESENCHYMAL STROMAL CELL CHARACTERISTICS: IMPLICATIONS FOR THEIR CLINICAL USE

Submitted by Helen Dimitriou

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INTRODUCTION: Bone marrow (BM) stroma represents a source of progenitor stromal cells, termed mesenchymal stromal cells (MSCs), which are multipotent and can differentiate into cartilage, bone, and adipose tissue. Several questions have arisen regarding their long-term expansion and their safety before use.

OBJECTIVE: Our goal was to assess the long-term expansion and safety of MSCs in clinical practice.

METHODS: MSCs from BM of children with benign hematologic disorders and solid tumors without BM involvement were isolated and cultured for 10 consecutive passages (P). Immunophenotypic and functional characteristics, apoptosis, and the expression of cell cycle regulatory genes (*p53*, *p16*, and *Rb*) and signal transduction genes (*H-Ras*) involved in oncogenesis were assessed.

RESULTS: MSCs expressed mesenchymal-related surface antigens, >85% from P1. They had the ability to differentiate into osteocytes, adipocytes, and chondrocytes (reverse-transcription polymerase chain reaction). Colony forming units (fibroblast) ranged from 40.71 ± 4.3 at P1 to 15.5 ± 6.7 at P10. Their doubling time was 2.01 ± 0.14 days at P1 and 3.5 ± 1.19 days at P9. A low

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