



Pediatric Neurology Part III: Chapter 137. Main steps of skeletal muscle development in the human: Morphological analysis and ultrastructural characteristics ... muscle (Handbook of Clinical Neurology)

Norma Beatriz Romero, Monica Mezmezian, Anna Fidzia?ska

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During embryogenesis, paraxial mesoderm undergoes segmentation into somites, progressing from head to tail. Somites differentiate into dermomyotomes, then into dermatomes and myotomes. Some head muscles derive from the anterior paraxial and precordial mesoderm. Between 10 and 13 weeks of gestation, the fusion of myoblasts generates primary myotubes with central nuclei, and the latter form the second generation of myotubes which requires active innervation. Nicotinamide adenine dinucleotide dehydrogenase-tetrazolium reductase appears before succinate dehydrogenase, and ATPase shows an intermediary activity. β -Sarcoglycan appears by 7 gestational weeks and α -sarcoglycan by 10–12 weeks. β -Spectrin, dystrophin, and utrophin appear by 9 weeks, vimentin and desmin appear by 10 weeks and stain strongly between 10 and 15 weeks. Slow, embryonic myosin heavy chain (MHC) isoenzymes appear before 15 weeks, whereas fetal fast MCH occur later (15–18 weeks). Myotubes become myofibers with peripheral nuclei between 15 and 18 weeks. Large muscle fibers (Wohlfart B) are visible by 20–21 weeks, Wohlfart A by 21–25 weeks. Perimysium surrounds compacted and grouped fibers by 24 weeks, and utrophin disappears, whereas dystrophin stains intensely. At 29 weeks, type I fibers are visible, and by 31–33 weeks they are mature for ATPase staining. Three types of type II fibers can be seen. Vimentin disappears between 15 and 30 weeks, while desmin remains weakly positive at birth.

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