



**Cover:** Immunostaining shows the generation of spinal motor neurons from human iPSCs at day 24 after differentiation. HB9<sup>+</sup> spinal motor neurons (red colour) were also positive for Tau protein (green colour). Blue indicates Hoechst-stained nuclei. See article by Xu et al. on page 39.

### CLINICAL PUZZLE

- 1 The role of enterocyte defects in the pathogenesis of congenital diarrheal disorders  
**Overeem, A. W., Posovszky, C., Rings, E. H. M. M., Giepmans, B. N. G. and van IJzendoorn, S. C. D.**

### RESEARCH ARTICLES

- 13 X-linked Christianson syndrome: heterozygous female *Slc9a6* knockout mice develop mosaic neuropathological changes and related behavioral abnormalities  
**Sikora, J., Leddy, J., Gulinello, M. and Walkley, S. U.**
- 25 The Menkes and Wilson disease genes counteract in copper toxicosis in Labrador retrievers: a new canine model for copper-metabolism disorders  
**Fieten, H., Gill, Y., Martin, A. J., Concilli, M., Dirksen, K., van Steenbeek, F. G., Spee, B., van den Ingh, T. S. G. A. M., Martens, E. C. C. P., Festa, P., Chesi, G., van de Sluis, B., Houwen, R. H. J. H., Watson, A. L., Aulchenko, Y. S., Hodgkinson, V. L., Zhu, S., Petris, M. J., Polishchuk, R. S., Leegwater, P. A. J. and Rothuizen, J.**
- 39 Abnormal mitochondrial transport and morphology as early pathological changes in human models of spinal muscular atrophy  
**Xu, C.-C., Denton, K. R., Wang, Z.-B., Zhang, X. and Li, X.-J.**

- 51 The cannabinoid CB<sub>1</sub> receptor and mTORC1 signalling pathways interact to modulate glucose homeostasis in mice  
**Bermudez-Silva, F. J., Romero-Zerbo, S. Y., Haissaguerre, M., Ruz-Maldonado, I., Lhamyani, S., El Bekay, R., Tabarin, A., Marsicano, G. and Cota, D.**

- 63 Effect of bromocriptine alginate nanocomposite (BANC) on a transgenic *Drosophila* model of Parkinson's disease  
**Siddique, Y. H., Khan, W., Fatima, A., Jyoti, S., Khanam, S., Naz, F., Rahul, Ali, F., Singh, B. R. and Naqvi, A. H.**

- 69 A new model for non-typeable *Haemophilus influenzae* middle ear infection in the *Junbo* mutant mouse  
**Hood, D., Moxon, R., Purnell, T., Richter, C., Williams, D., Azar, A., Crompton, M., Wells, S., Fray, M., Brown, S. D. M. and Cheeseman, M. T.**

### RESOURCE ARTICLES

- 81 Urine-sample-derived human induced pluripotent stem cells as a model to study PCSK9-mediated autosomal dominant hypercholesterolemia  
**Si-Tayeb, K., Idriss, S., Champon, B., Caillaud, A., Pichelin, M., Arnaud, L., Lemarchand, P., Le May, C., Zibara, K. and Cariou, B.**
- 91 Longitudinal micro-CT provides biomarkers of lung disease that can be used to assess the effect of therapy in preclinical mouse models, and reveal compensatory changes in lung volume  
**Vande Velde, G., Poelmans, J., De Langhe, E., Hillen, A., Vanoirbeek, J., Himmelreich, U. and Lories, R. J.**