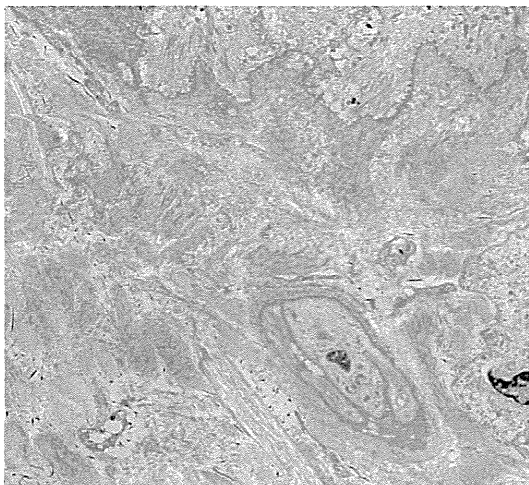


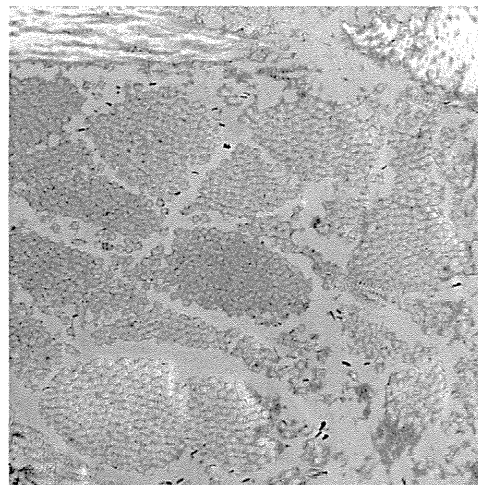
図 2

図2a: 電顕所見 (Patient 12)

単純電顕

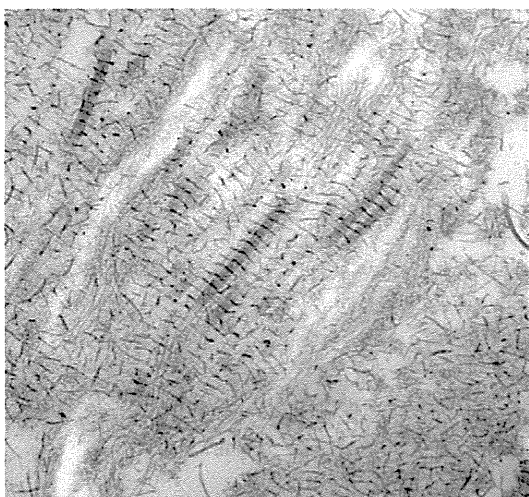


真皮表層 (UAのみ)

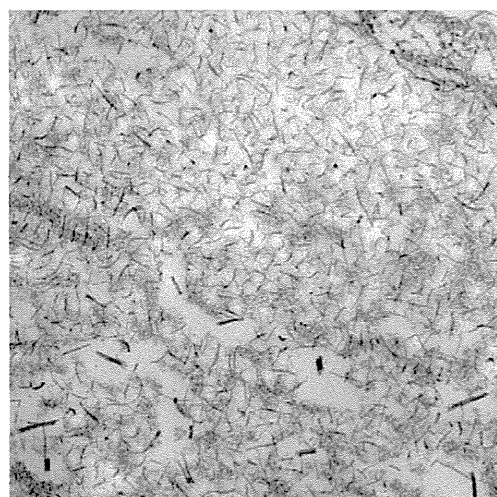


真皮深層 (UAのみ)

真皮表層

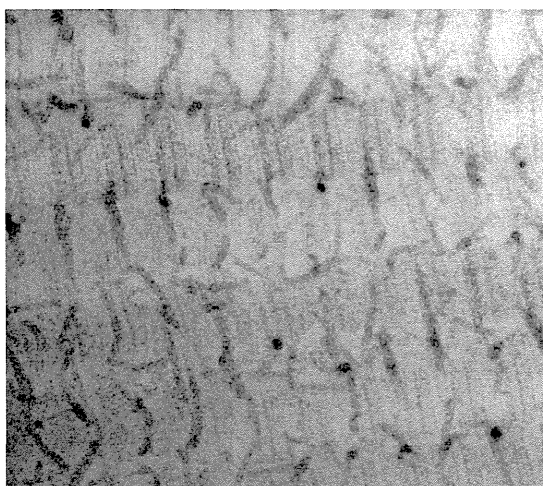


CB+UA (縦断像)

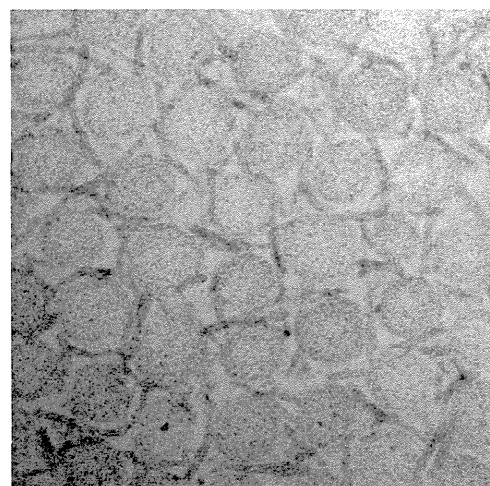


CB+UA (横断像)

真皮深層



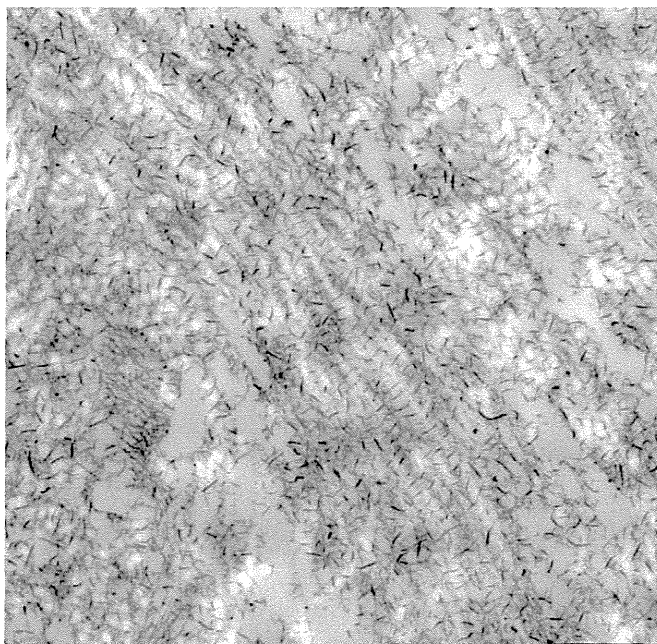
CB+UA (縦断像)



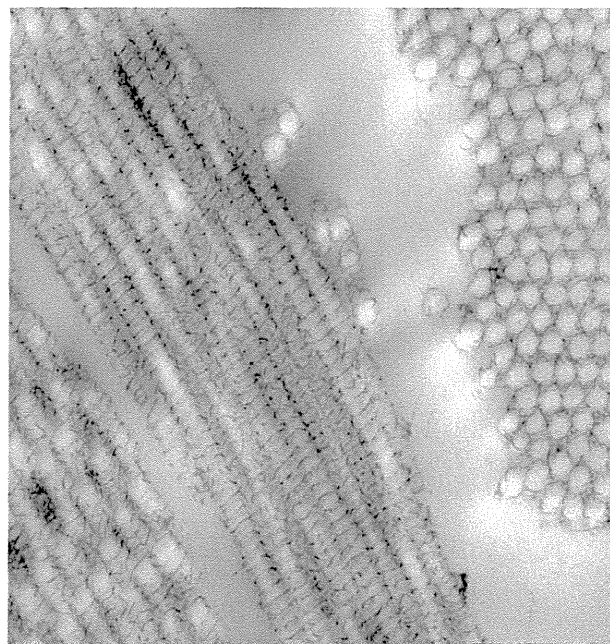
CB+UA (横断像)

図2b: 電顕所見 (Patients 21、30)

Patients 21

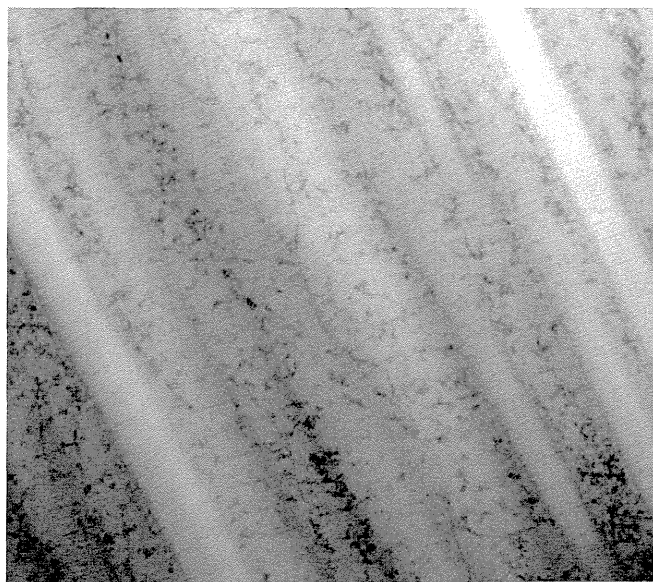


真皮表層:CB+UA

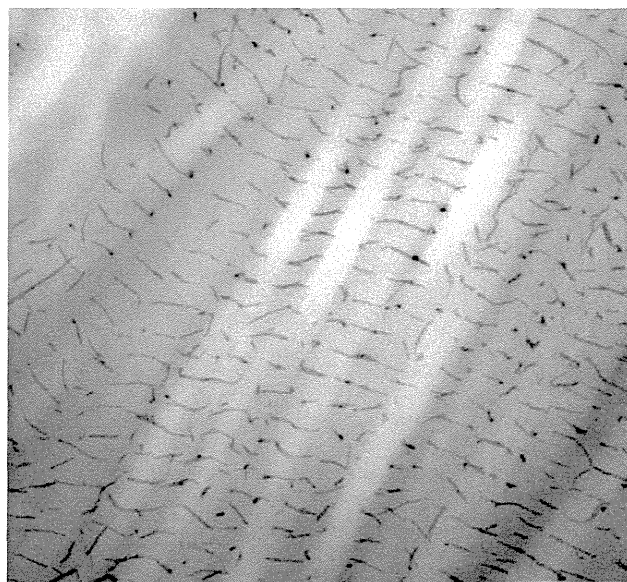


真皮深層:CB+UA

Patients 30

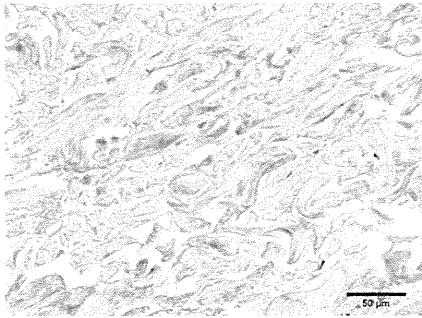


真皮深層:UAのみ

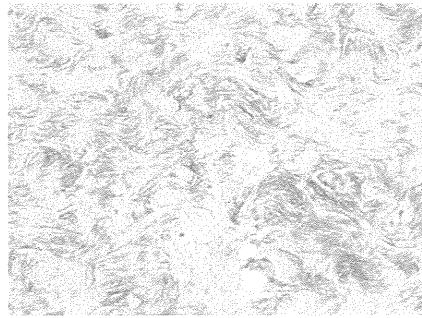


真皮深層:CBのみ

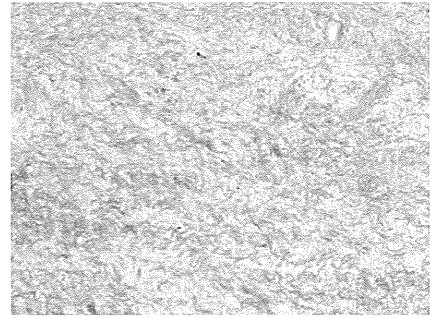
患者皮膚を用いた抗デコリン免疫組織化学解析



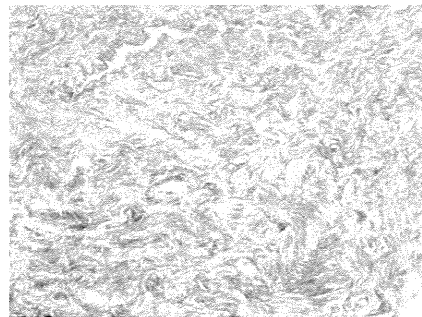
健常14歳女兒、上腕



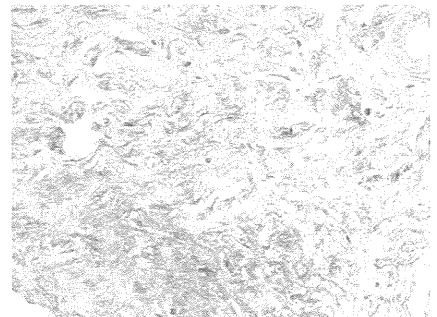
DDEDS16歳女兒、腹部



DDEDS19歳女性、上腕



DDEDS18歳女性、背部



DDEDS4歳女兒、上腕

图 4

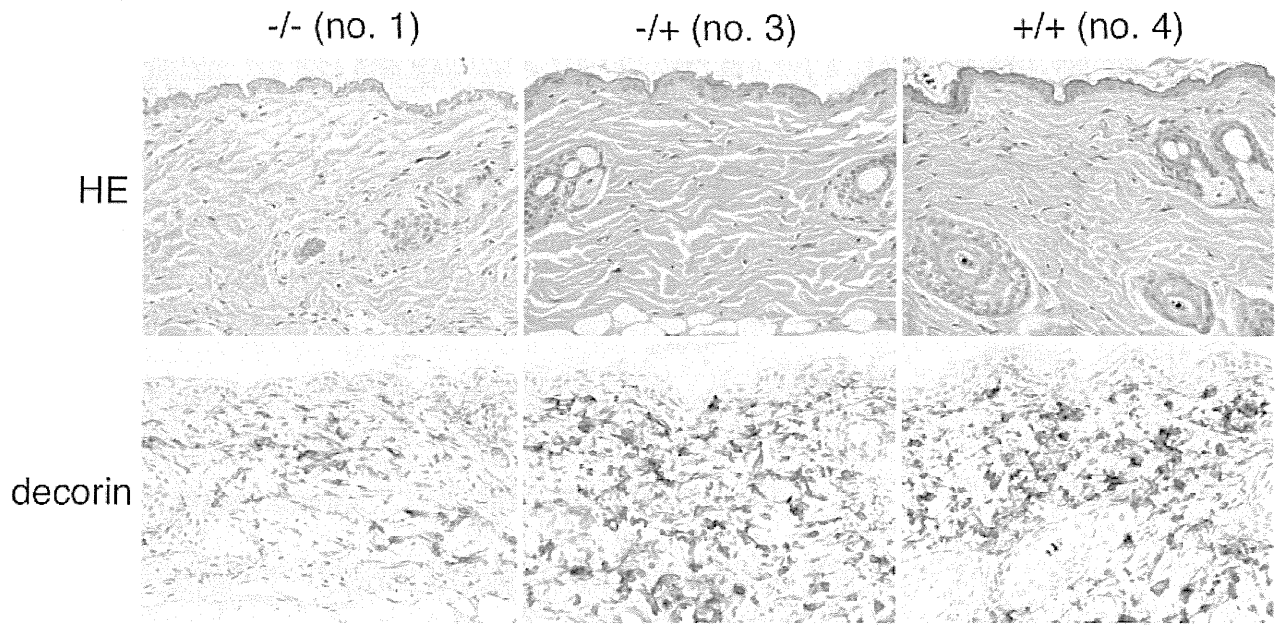


图 5

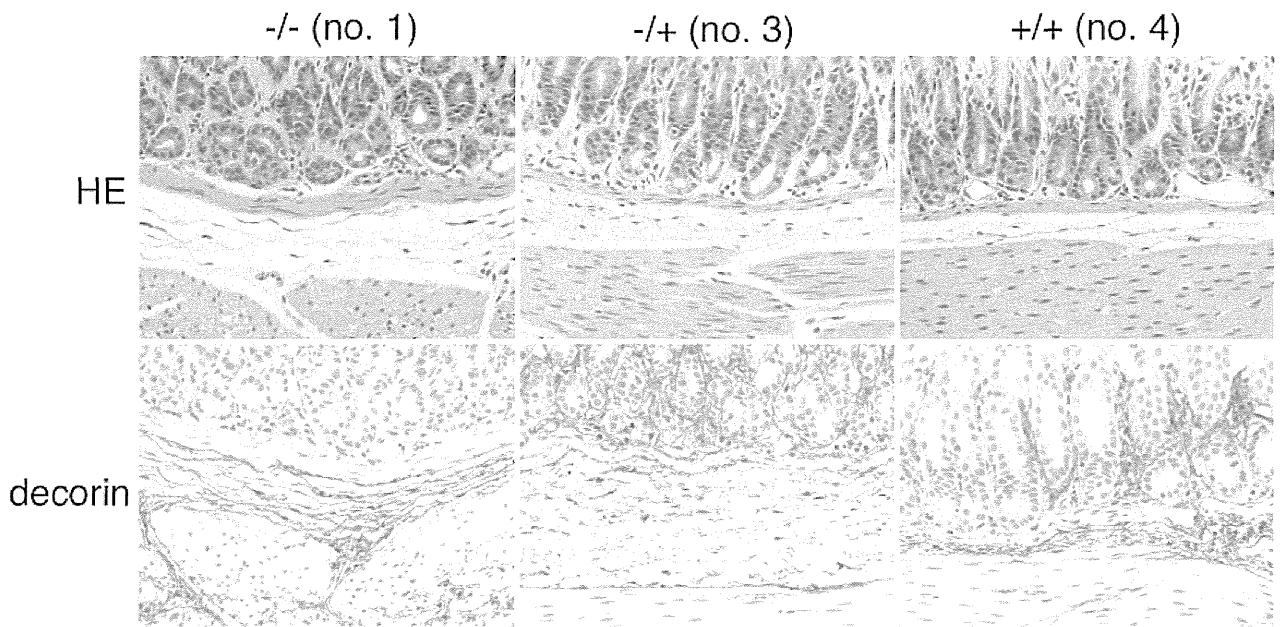
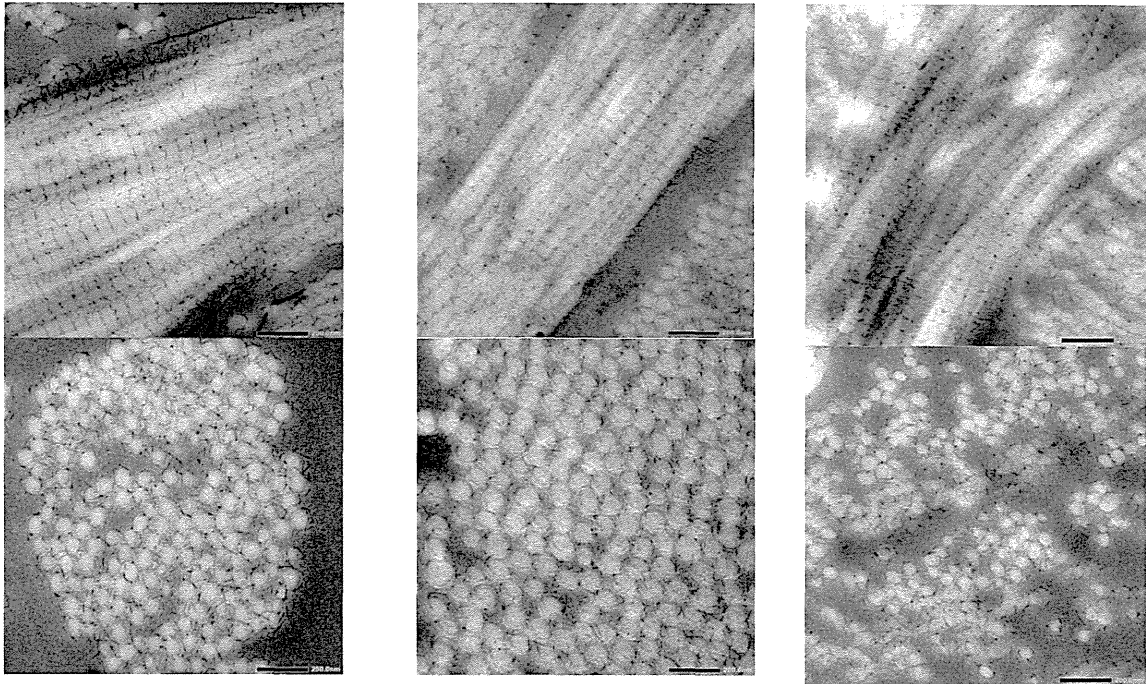


图 6

皮膚表層



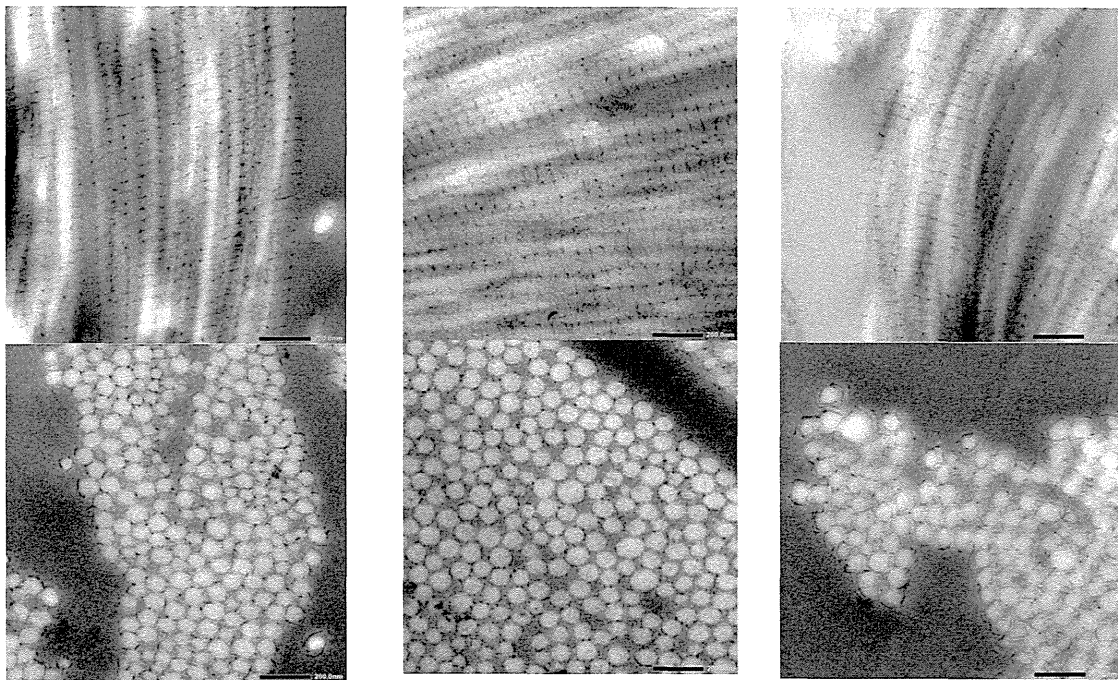
WT

Chst14^{+/-}

Chst14^{-/-}

图 7

皮膚深層

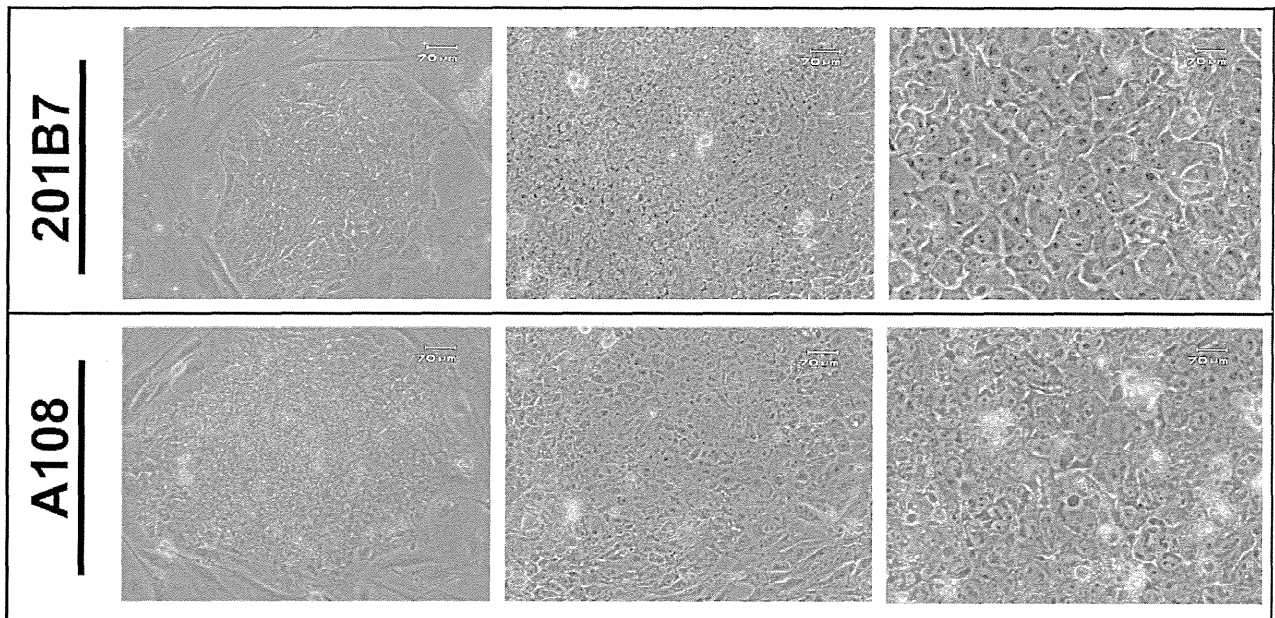


WT

Chst14^{+/-}

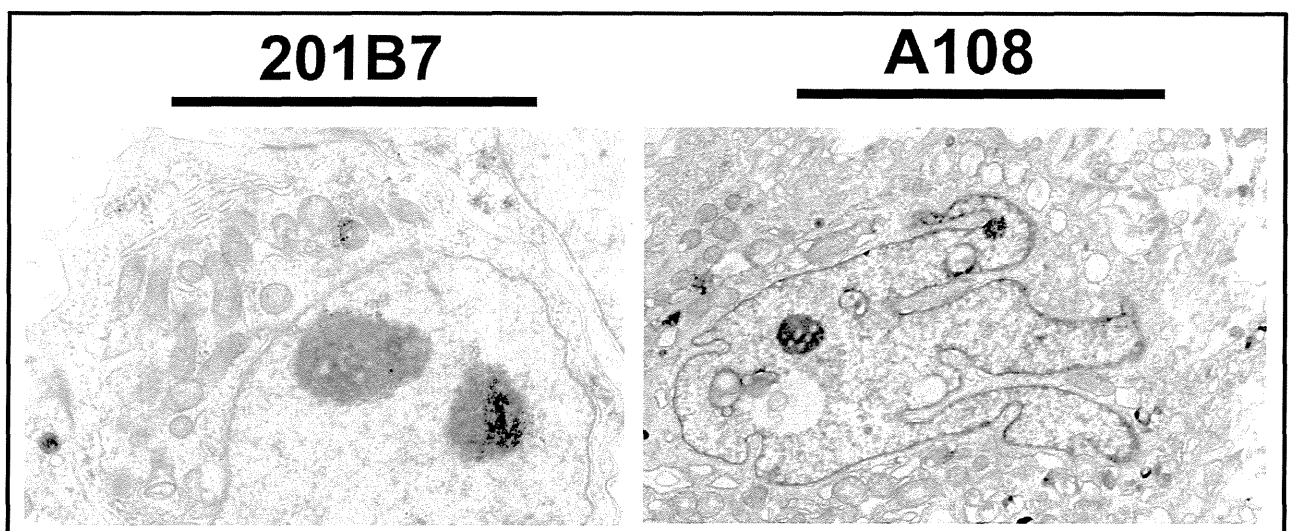
Chst14^{-/-}

☒ 8

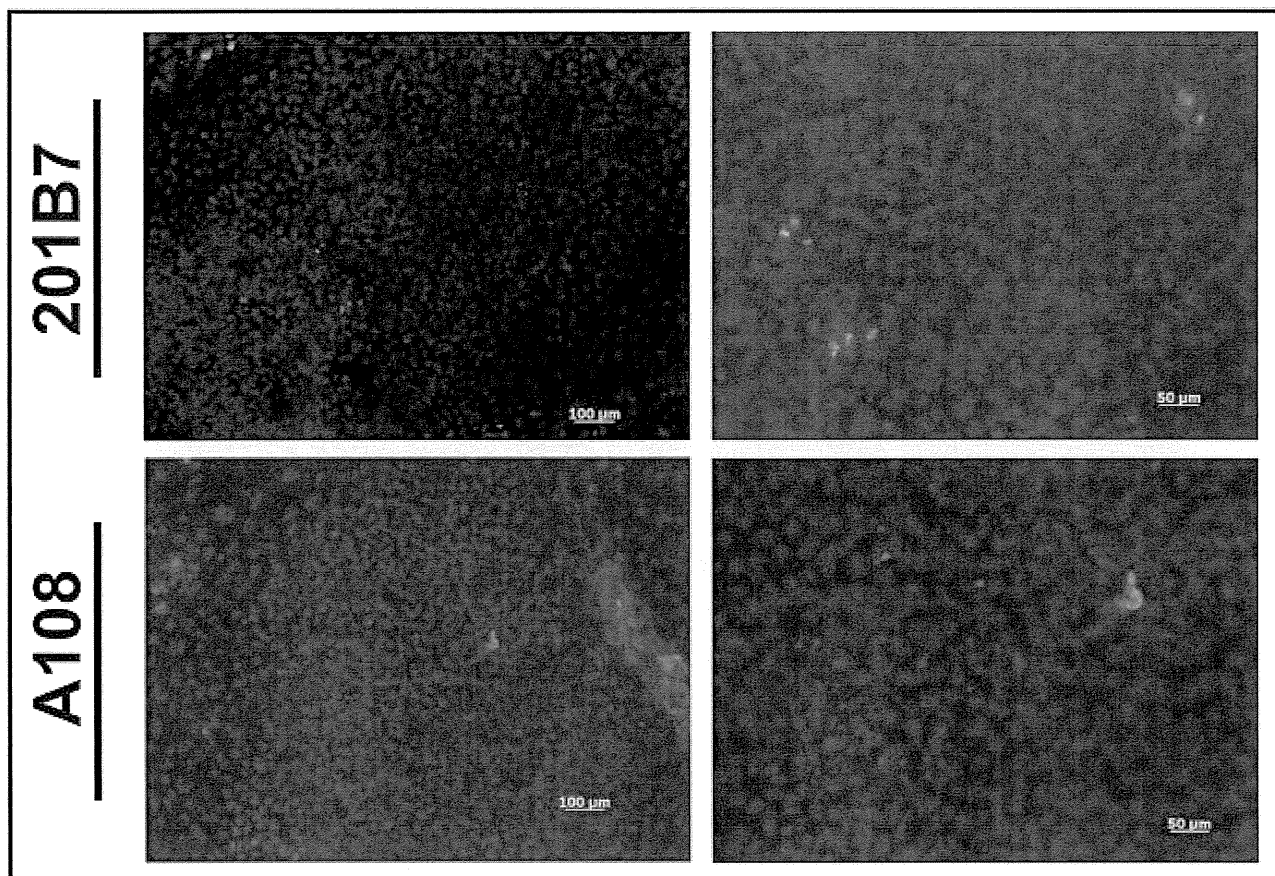


Undifferentiated iPS cells grown on mouse embryonic fibroblasts (MEF) were tightly packed in the colony and had a high nucleus-to-cytoplasm ratio. However, some differences were shown between patient-iPS and normal iPS: (i) the size of A108 is smaller than 201B7; (ii) the boundary of A108 is obscure, even some cells were fused; (iii) some nuclei of A108 is different from that of 201B7, and more vacuoles were found in A108.

☒ 9

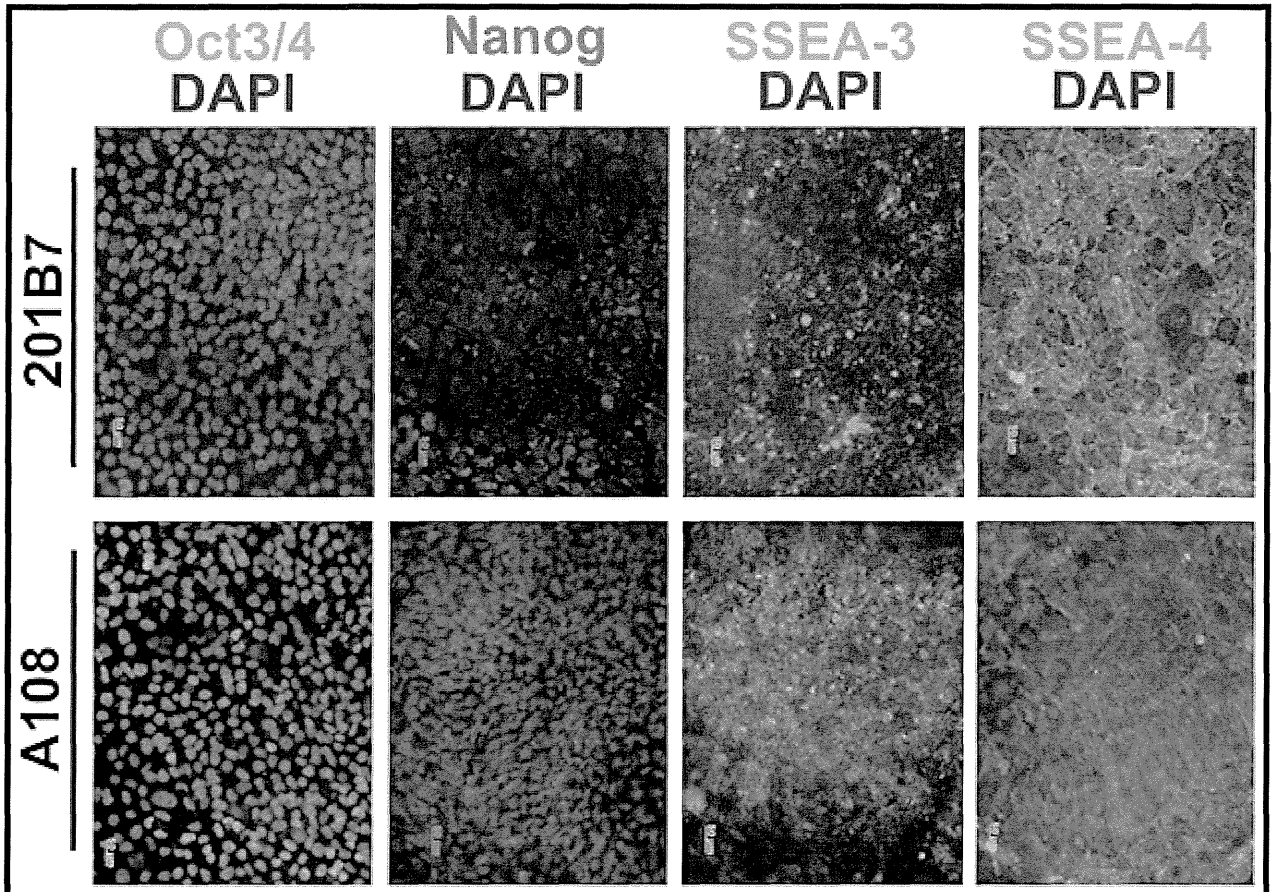


Lots of vacuoles were found in A108.



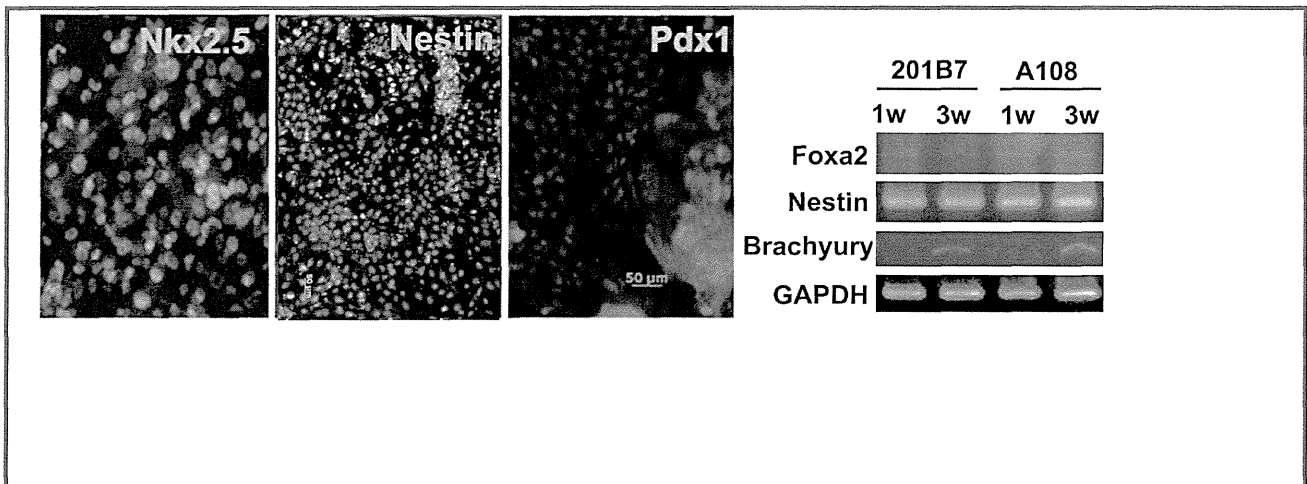
No significant difference of apoptosis was shown between A108 and 201B7.

图 11



The colonies expressed high levels of undifferentiated cell markers such as Oct3/4, Nanog, SSEA-3 and SSEA-4.

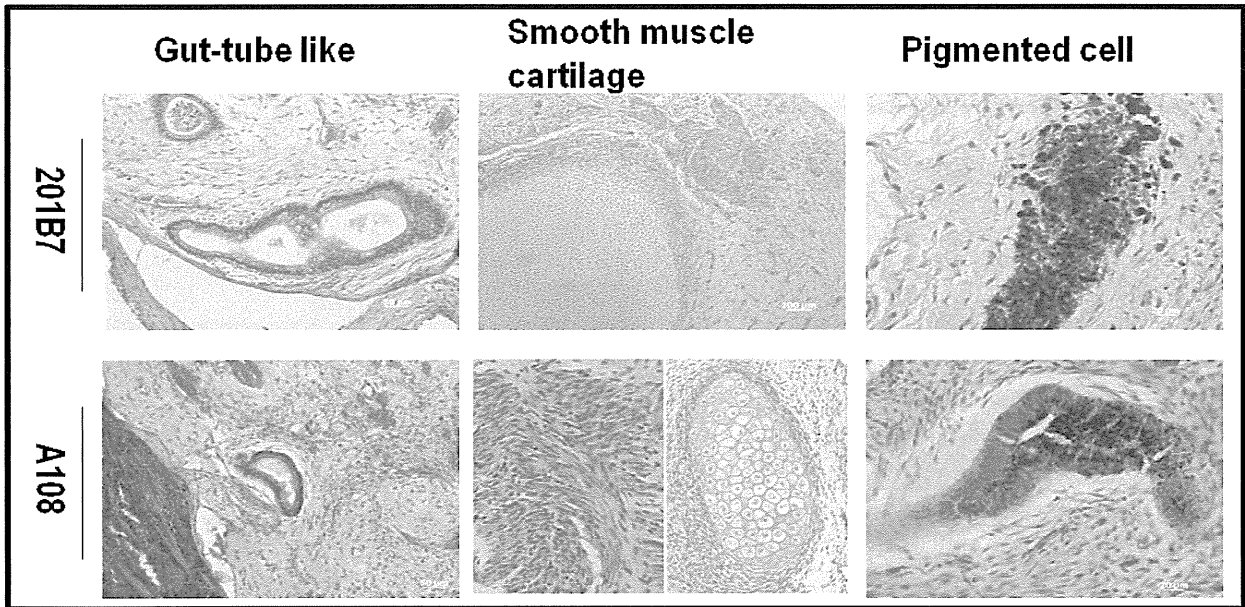
图 12



Expression of germ layer markers in EB outgrowths.

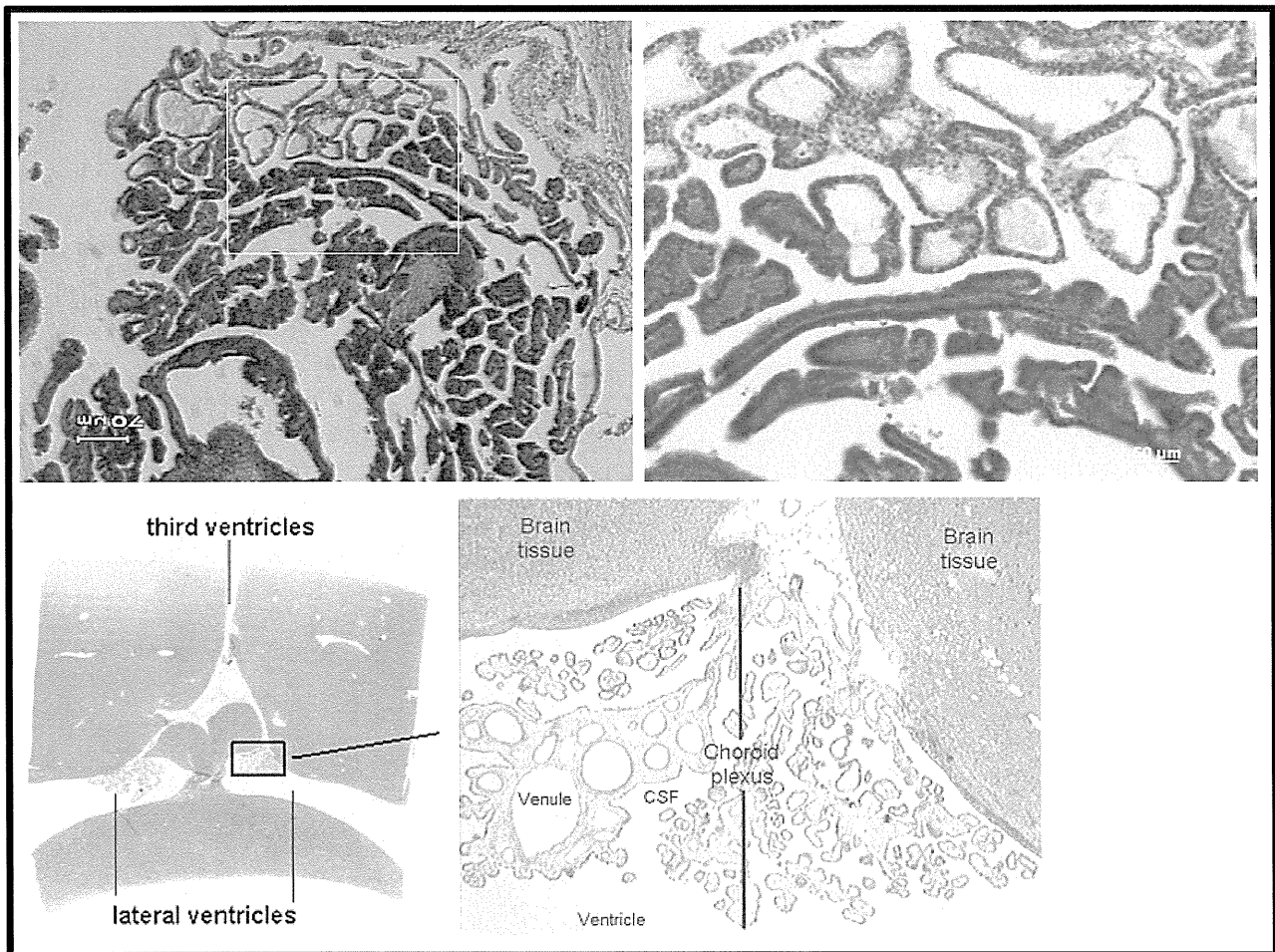
Endoderm: Foxa2, Pdx1; Ectoderm: Nestin; Mesoderm: Nkx2.5, Brachyury

☒ 13

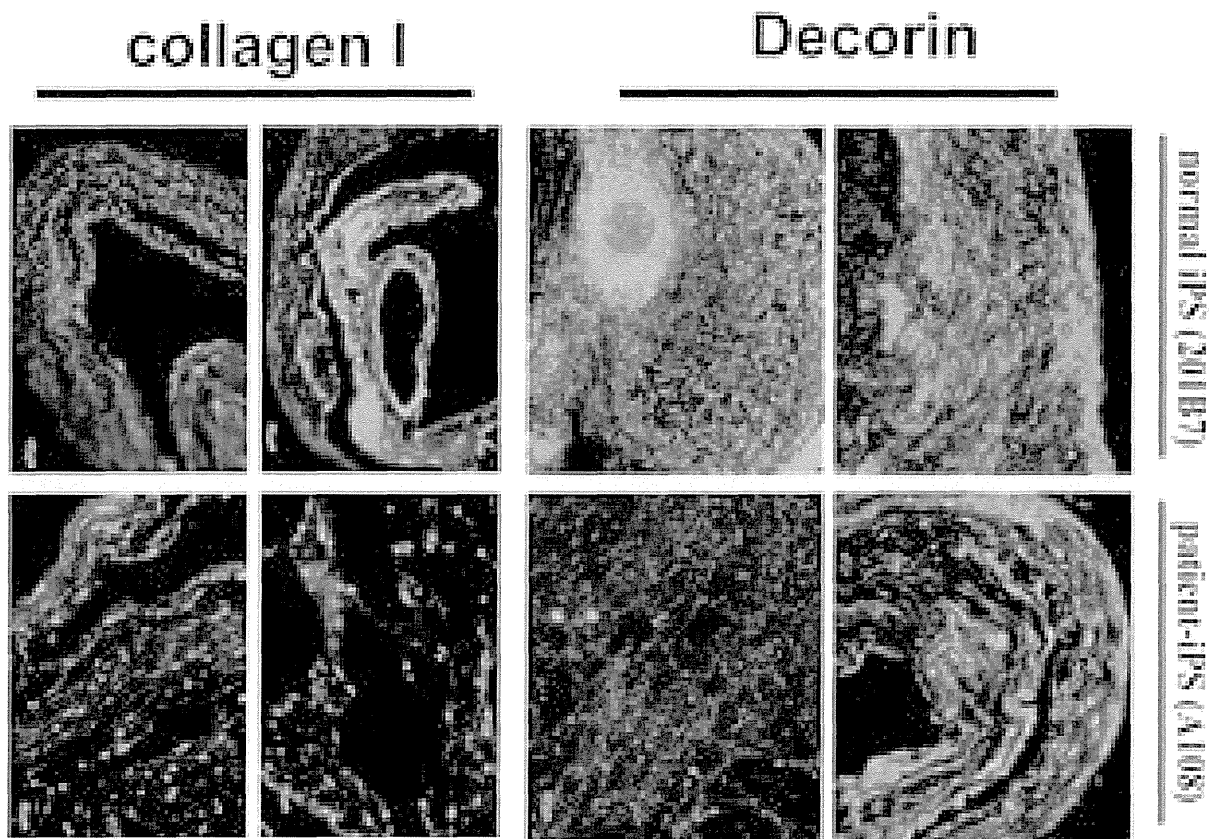


Teratoma was formed within 4 weeks after the transplantation of iPS. Tissues from three germ layer were found in teratoma.

☒ 14



Choroid plexus-like structure was found in teratoma formed from DD-EDS derived iPS.



Decorin and collagen I fiber in DD-EDS iPS-derived teratoma is slimmer than that in normal iPS-derived teratoma.