PIPADOR REVISION

MRCOG Part 1 Revision Notes

Neural Crest Derivatives

EMBRYOLOGY • LECTURE 50

VISUAL EXAMINATION REVISION NOTES

2026 Edition

MRCOG Part 1 Revision Notes

Lecture 50: Neural Crest Derivatives

Embryology • Visual revision edition

COLOUR CODE USED THROUGHOUT THESE NOTES

NEURAL CREST	CRANIAL	CARDIAC	VAGAL / ENTERIC	TRUNK

LEARNING OBJECTIVES — tick off as you go

- ☐ Describe how neural crest forms, delaminates and migrates.
- ☐ Name the functional regions and their main derivatives.
- ☐ List neural crest derivatives by system, including the pharyngeal arches.
- ☐ Explain neurocristopathies: cardiac, enteric, pigment, craniofacial and tumours.
- ☐ Apply to 22q11.2 deletion, Hirschsprung disease, phaeochromocytoma and melanoma in pregnancy.

1 Formation and Migration

IN ONE LINE: *A transient, multipotent, migratory population from the neural plate border — sometimes called the 'fourth germ layer'.*

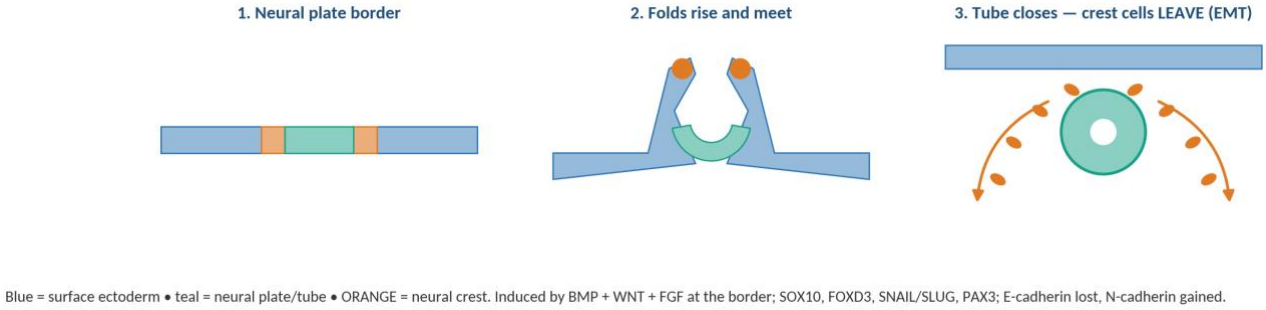


Figure 1. From neural plate border to migrating cells.

Step	Key molecules
Induction	BMP, WNT, FGF at the neural plate border
Specification	PAX3, SNAIL/SLUG, FOXD3, SOX9, SOX10
Delamination (EMT)	Loss of E-cadherin, gain of N-cadherin, matrix metalloproteinases
Migration	Ephrins, semaphorins, extracellular matrix cues

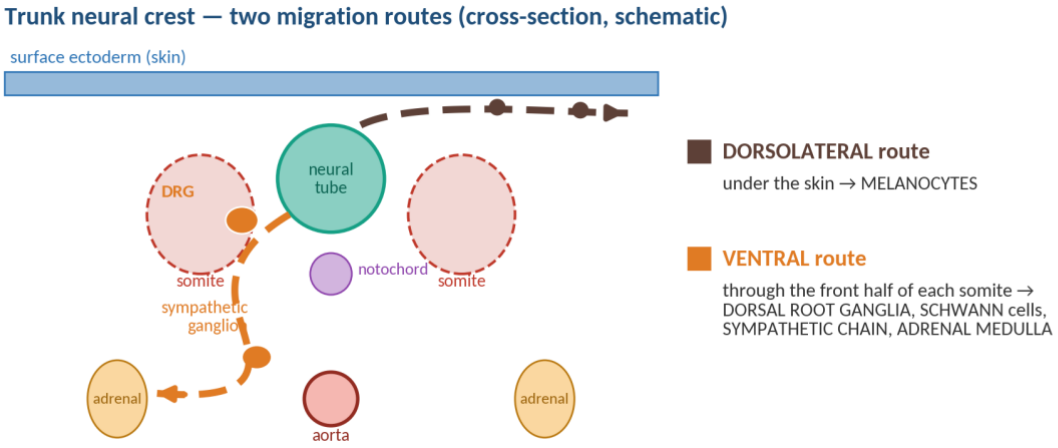


Figure 2. Trunk crest: two routes.

2 Functional Regions

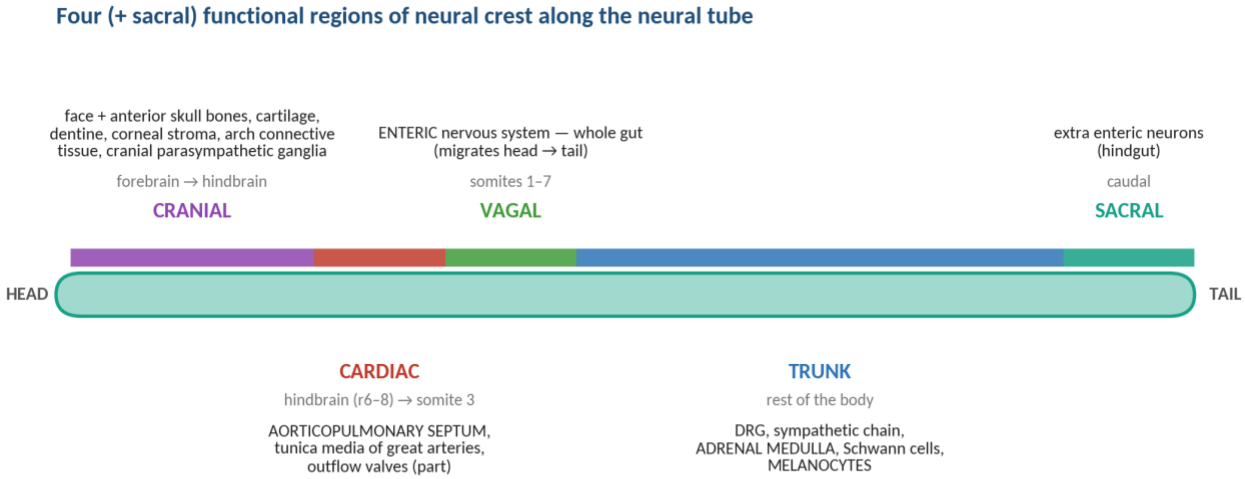


Figure 3. Regions along the neural tube.

3 Derivatives by System

What neural crest makes

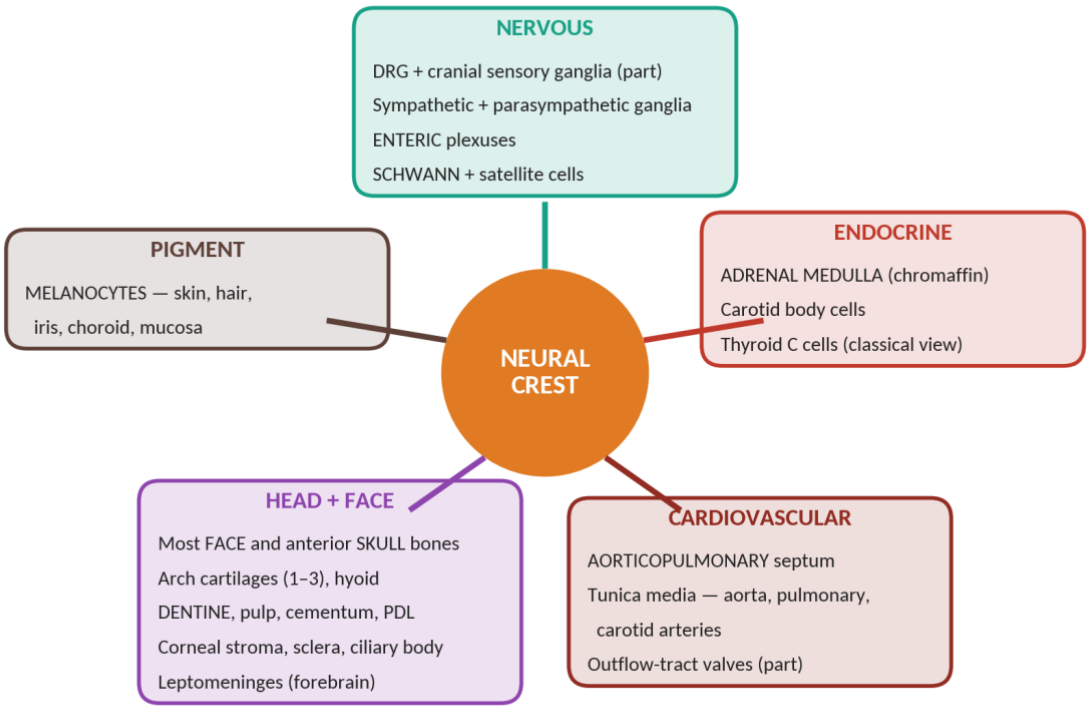


Figure 4. Neural crest derivatives.

- **Thyroid C cells** reach the thyroid in the ultimobranchial body (pouch 4); classically neural crest, though recent lineage studies suggest an endodermal origin (as in Lecture 49).
- **Face and anterior skull**: frontal, nasal, parts of temporal, maxilla, mandible (membrane bone), hyoid; the posterior skull base (chondrocranium) is mesodermal.

MNEMONIC / MEMORY HOOK

MOTEL PASS (corrected): **M**elanocytes • **O**dontoblasts (dentine) • **T**unica media of great arteries • **E**nteric nervous system • **L**eptomeninges (forebrain) • **P**arafollicular C cells (classical) • **A**drenal medulla + **A**ll PNS ganglia • **S**chwann cells • **S**piral (aorticopulmonary) septum. Plus most **skull and face bones**.

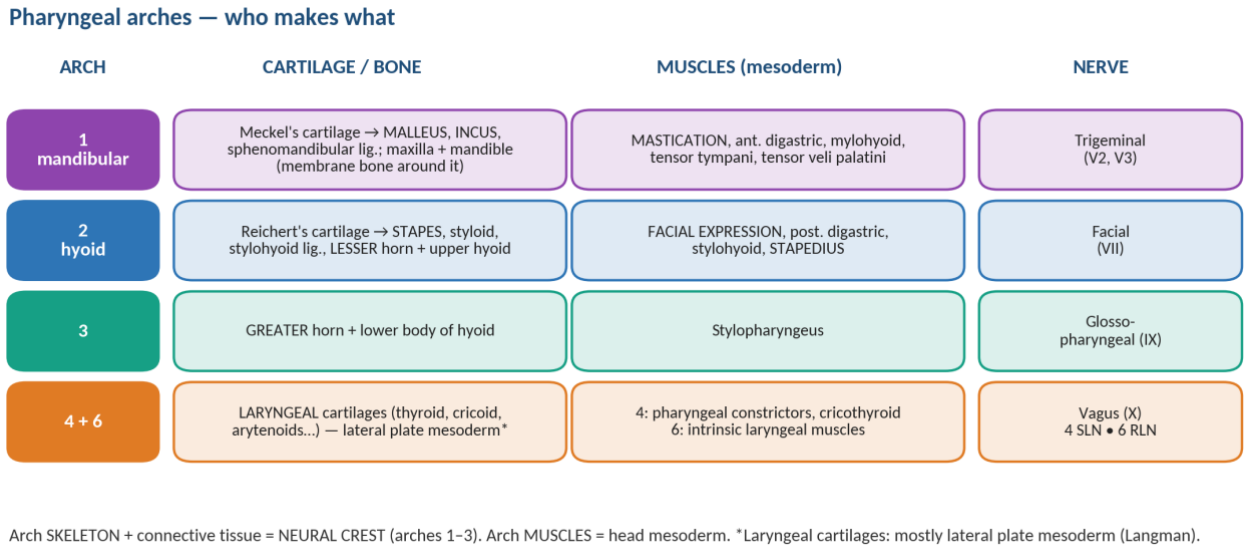


Figure 5. Pharyngeal arches.

4 Neurocristopathies

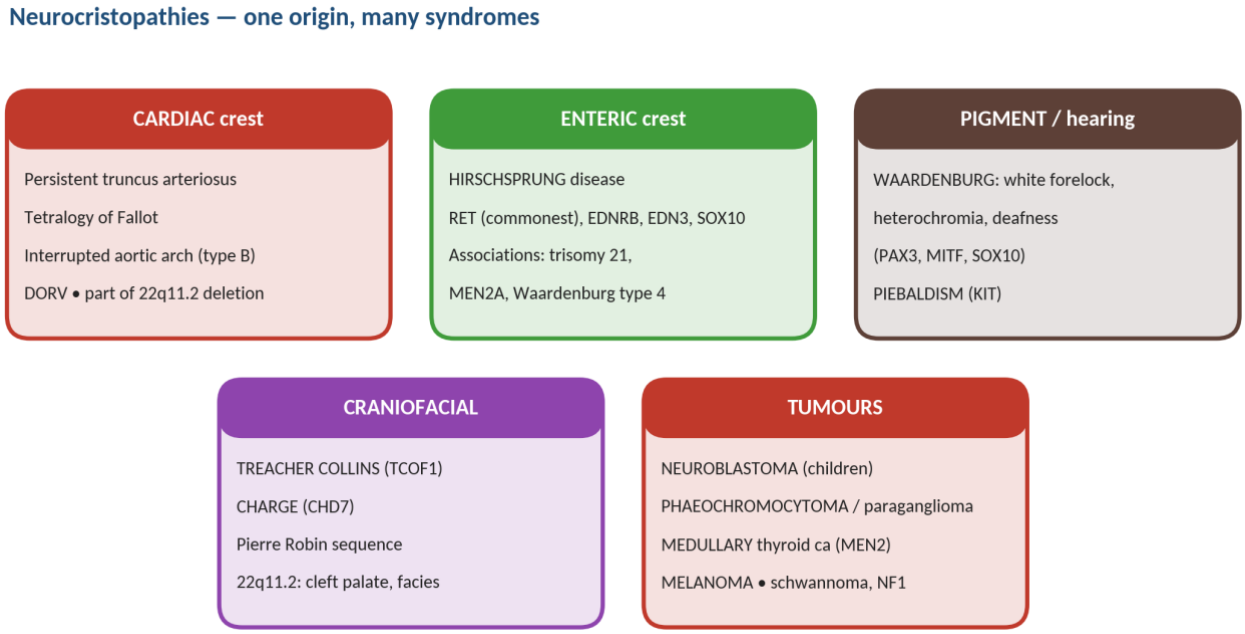


Figure 6. One origin, many syndromes.

22q11.2 DELETION (DiGeorge)

- **Cardiac crest + pouches 3/4**
- **Conotruncal** defects: tetralogy, interrupted arch (type B), truncus
- Thymic hypoplasia (T-cell deficiency), **hypoparathyroidism (hypocalcaemia)**
- Cleft palate, facial features, learning difficulty

HIRSCHSPRUNG DISEASE

- Crest fails to reach the distal hindgut → **aganglionic segment, always including the rectum**
- Delayed meconium (> 24–48 h), distension, constipation
- **Rectal suction biopsy**: no ganglion cells, ↑ acetylcholinesterase nerve fibres (calretinin lost)
- Genes: **RET**, EDNRB, EDN3, SOX10 • trisomy 21, MEN2A, Waardenburg 4

PHAEOCHROMOCYTOMA

- Adrenal-medulla (chromaffin) tumour — headache, sweating, palpitations, hypertension
- The old 'rule of 10s' is **outdated**: **~30–40% are hereditary** (MEN2, VHL, NF1, SDHx)
- In pregnancy: rare but high mortality if missed — alpha-blockade before beta-blockade

MELANOMA IN PREGNANCY

- One of the commoner cancers in pregnancy (with breast, cervical cancer, lymphoma)
- The tumour **most likely to spread to the placenta and fetus** (still rare)
- Send the **placenta for histology**; examine the baby

- Others: **Waardenburg** (PAX3, MITF, SOX10), **piebaldism** (KIT), **NF1**, **MEN2A/2B** (RET: medullary thyroid cancer, phaeochromocytoma), **CHARGE** (CHD7), **Treacher Collins** (TCOF1), Pierre Robin sequence, **neuroblastoma**.

★ **PEARL**

Cardiac crest builds the aorticopulmonary septum — so crest defects give **conotruncal** anomalies (truncus, tetralogy, interrupted arch), classically with **22q11.2 deletion**.

Hirschsprung = crest failing to colonise the gut, always involving the rectum; **RET** links it to **MEN2**.

5 MRCOG Exam Pearls — Last-Minute Page

Numbers to know

Wk 3–4 Crest forms (neurulation)	**4 (+1)** Regions (+ sacral)	**2** Trunk migration routes	**1–3** Arches with crest skeleton
~30–40% Hereditary phaeos	**22q11.2** DiGeorge deletion	**> 24–48 h** Delayed meconium	**RET** Hirschsprung + MEN2 gene

HIGH-YIELD CHECKLIST

- ☐ From the neural plate border; EMT (E- → N-cadherin); SOX10, FOXD3, PAX3.
- ☐ Trunk routes: dorsolateral → melanocytes; ventral → DRG, sympathetic chain, adrenal medulla, Schwann cells.
- ☐ Cranial → face/skull bones, dentine, corneal stroma; cardiac → AP septum; vagal/sacral → enteric NS.
- ☐ Arch skeleton (1–3) = crest; arch muscles = mesoderm; laryngeal cartilages mostly mesoderm.
- ☐ Enamel ectoderm vs dentine crest.
- ☐ 22q11.2: conotruncal + thymus + parathyroids.
- ☐ Hirschsprung: RET; always rectum; suction biopsy.
- ☐ Tumours: neuroblastoma, phaeochromocytoma, MTC, melanoma, schwannoma.
- ☐ Waardenburg (PAX3, MITF, SOX10); Treacher Collins (TCOF1); CHARGE (CHD7).

Self-assessment — cover the right-hand column

Question	Answer
Where does neural crest arise?	At the border of the neural plate and surface ectoderm
Which crest region builds the aorticopulmonary septum?	Cardiac crest
Which crest colonises the gut?	Vagal (whole gut) + sacral (hindgut)

Question	Answer
Crest cells taking the dorsolateral route become…?	Melanocytes
Origin of dentine vs enamel?	Dentine — crest; enamel — ectoderm
Arch 2 skeletal derivatives?	Stapes, styloid, lesser horn + upper hyoid
Main gene in Hirschsprung disease?	RET
Three features of 22q11.2 deletion?	Conotruncal heart defect, hypocalcaemia, immune deficiency
Five crest tumours?	Neuroblastoma, phaeo, medullary thyroid ca, melanoma, schwannoma
Cancer most likely to reach the placenta/fetus?	Melanoma

6 References

- Sadler TW. Langman's Medical Embryology. 14th ed. 2019.
- Lenders JWM et al. Pheochromocytoma and paraganglioma: Endocrine Society guideline. 2014.
- Moore KL, Persaud TVN, Torchia MG. The Developing Human. 11th ed. 2020.