

# BÖLÜM 17

## ÖN BEYİN GELİŞİMSEL BOZUKLUKLARI

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### GİRİŞ

Serebral korteksin gelişimi, birkaç gestasyonel hafta içinde ortaya çıkan karışık ve dinamik bir süreçtir.<sup>1</sup> Gestasyonun 3-5. haftası arasında nöral tüpün rostralinde 3 ana vezikül oluşur; prozensefalon (telensefalon ve diensefalon), mezensefalon (midbrain) ve rombensefalon (metencephalon ve myelensefalon). Telensefalondan putamen, serebral hemisferler ve nükleus kaudatus oluşurken; diensefalondan talamus, hipotalamus, glopus pallidum ve optik veziküller oluşur.<sup>2</sup>

Gelişimsel bozukluklar ve epilepside görüntüleme yöntemlerindeki ilerlemeyle birlikte gittikçe artan oranda anormal kortikal gelişim saptanmaktadır. Prozensefalik oluşumdaki başlıca bozukluklar holoprozensefali, korpus kallozum agenezisi ve septo-optik displazidir.

### Prozensefalonun Normal Gelişimi

Prozensefalonun gelişimi; prozensefalik formasyon, prozensefalik bölünme ve orta hat prozensefalik gelişimi şeklinde üç ardışık olayla gerçekleşir (Tablo 1). Gestasyonun yaklaşık olarak 5.haftasında prozensefalon, telensefalon ve diensefalona bölünür. Telensefalondan putamen ve serebral hemisferler, oluşurken; dien-

sefalondan ise talamus, hipotalamus, glopus pallidus ve optik veziküller oluşur.<sup>2</sup>

Postkonsepsiyonel 35. günlerde karmaşık sinyal yollarının kontrolü altında, artmış apoptozis ve azalmış proliferasyon sonucu telensefalon sağ ve sol veziküllere bölünür. Bu ilişkilerdeki bozukluklar holoprozensefali anomaliler spektrumuna yol açabilir.<sup>3</sup> İnter-hemisferik boşlukta bulunan spesifik klavuz ve büyüme faktörlerinin etkisi altında kallozal liflerden, 14-15. haftalarda matur korpus kallozumun beş parçası (rostrum, genu, body, isthmus, splenium) kısa da olsa hızlıca gelişir. Korpus kallozum 18-22. haftalar arasında tam uzunluğuna ulaşır ve ardından intrauterin ve infantil dönem boyunca kalınlaşır.<sup>4,5</sup>

**Tablo 1: Prozensefalik Gelişim**

Prozensefalik formasyon
Prozensefalik bölünme
• Optik ve olfaktor yapılar
• Telensefalon → Serebral hemisferler
• Diensefalon → Talamus, hipotalamus
Orta hat prozensefalik gelişim
• Orta hat prozensefalik gelişim
• Korpus kallosun, septun pellucidum, optik sinirler (kiazm), hipotalamus

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