

# İMMÜNGLOBULİN-A VASKÜLİTİ (HENOCH- SCHONLEIN PURPURASI)

## 7. BÖLÜM

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### GİRİŞ

Henoch-Schönlein Purpurası (HSP); tarihsel olarak ilk kez 1801 yılında Heberden tarafından bildirilmiş olup, 1837'de Schönlein tarafından hastalığın purpura ve artrit bulguları, 1874 ve 1899'da ise Henoch tarafından sırası ile gastrointestinal ve renal tutulumu bildirilmiştir. İmmünglobulin A vaskülit (IgA vaskülit) [IgAV]; eski adı ile HSP)<sup>(1)</sup>, çocuklarda en sık görülen sistemik vaskülit formudur. Vakaların %90'ı çocuk yaş grubunda görülür. Diğer birçok sistemik vaskülit formunun aksine IgAV (HSP), vakaların büyük çoğunluğunda kendi kendini sınırlar. Hastalık kendini; trombositopeni ve koagülopati olmadan palpable purpura, artrit/artralji, karın ağrısı ve renal tutulum tetradı ile karakterize eder.

### EPİDEMİYOLOJİ

IgAV (HSP) primer olarak 3 ile 15 yaş arasında ortaya çıkan bir çocukluk çağı hastalığıdır<sup>(2)</sup>. İngiltere'de yapılan bir çalışmada 17 yaş altı çocuklarda yıllık insidansın 100.000'de 20 olduğu; 4-6 yaş arasındaki çocuklarda 100.000'de 70 oranında bir pik insidansının olduğu tespit edilmiştir<sup>(3)</sup>. Tayvan ve Çek Cumhuriyeti'nde yapılan çalışmalarda ise 17 yaş altı çocuklarda 100.000'de 10'luk daha düşük bir insidans saptanmış ve pik insidansın 5-7 yaş arasında olduğu bildirilmiştir<sup>(4,5)</sup>. IgAV (HSP) yetişkinlerde daha az sıklıkla görülmektedir<sup>(6)</sup>. İspanya ve Kore'de yapılan iki retrospektif çalışmada IgAV (HSP) hastalarının %25-30'u yetişkin olup; her iki kohortta da erişkinlerin çocuk hastalara göre önemli ölçüde daha kötü renal sonuçlarının olduğu gözlenmiştir<sup>(7,8)</sup>.

Çoğu çalışma erkek:kadın oranının 1,2:1 ile 1,8:1 oranlarında erkeklerde daha fazla görüldüğünü belirtmiştir<sup>(3,4,9-11)</sup>. IgAV (HSP), Asyalı çocuklarda daha sık ol-

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## SONUÇ

IgAV (HSP) çocuklarda en sık görülen sistemik vaskülit formudur. Altta yatan nedeni hala net olarak bilinmemektedir. Tanısı genellikle hastalığın klinik belirtileri ile konulmaktadır. Genellikle kendi kendini sınırlayan ve prognozu iyi olan bir hastalık olsa da küçük bir azınlıkta (<%1) başta böbrek hastalığı olmak üzere uzun süreli komplikasyonlar gelişebilmektedir. IgAV (HSP) gelişen hastalar, hastalığın kendisi, gelişebilecek komplikasyonları ve özellikle potansiyel olarak ilerleyici böbrek tutulumu açısından tetkik edilip düzenli takip edilmeli ve uygun şekilde tedavi edilmelidir.

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