Conjoined twins are one of the rarest and most challenging congenital anomalies in pediatric surgery. Successful surgical separation is difficult because it is technically typical to separate shared vital organs successfully. The timing of separation is variable, but separation is usually delayed until such infants are relatively mature (i.e., 9-12 months of age). Operative survival was 50% in the neonatal period, but 90% in those over 4 months of age. The present case was successfully separated early because of cardiac problems in one of the twins. These twins were omphalopagus and only the liver was shared through a bridge. The vascular and biliary trees were independent from each other. Successful surgical separation was on 11th day of life, but one died of sepsis 18 days after operation.

Index Words: Conjoined twins, Omphalopagus
Fig. 1. These twins lie face to face and are joined from the xiphoid to the umbilicus. Omphalocele is noticed.

Fig. 2. This barium study shows that intestinal tracts of each baby are not communicated.

Fig. 3. These twins have a bridge of hepatic tissue. Other organs are not communicated on MRI.

Examinal signs (Fig. 1) show the two babies have small umbilicus, and from the xiphoid to the umbilicus. Examination revealed that Omphaloceles are not communicated.

This barium study shows that intestinal tracts of each baby are not communicated (Fig. 2). Examination revealed that these twins have a bridge of hepatic tissue. Other organs are not communicated on MRI (Fig. 3).

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Table 1. Laboratory Findings

<table>
<thead>
<tr>
<th></th>
<th>I</th>
<th>II</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC (×10⁹/mm³)</td>
<td>11.4</td>
<td>11.7</td>
</tr>
<tr>
<td>Hg (g/dL)</td>
<td>15.8</td>
<td>14.2</td>
</tr>
<tr>
<td>Hct (%)</td>
<td>46.9</td>
<td>41.4</td>
</tr>
<tr>
<td>Platelet (×10⁹/mm³)</td>
<td>210</td>
<td>286</td>
</tr>
<tr>
<td>ALP (U/L)</td>
<td>137</td>
<td>149</td>
</tr>
<tr>
<td>AST (U/L)</td>
<td>130</td>
<td>45</td>
</tr>
<tr>
<td>ALT (U/L)</td>
<td>51</td>
<td>45</td>
</tr>
<tr>
<td>BUN (mg/dL)</td>
<td>3.0</td>
<td>4.2</td>
</tr>
<tr>
<td>Creatinine (mg/dL)</td>
<td>0.2</td>
<td>0.5</td>
</tr>
</tbody>
</table>

Fig. 4. This photograph shows complete separation of the twin.

결합생장이는 일반적으로 약 5만 내지 10만 분만
건수iazza 1건 정도의 빈도를 보이는 것으로 알려져
있으며1-3, 아프리카에서는 14,000 건당 1건으로
혼인에서 그 빈도가 약간 높은 것으로 보고되고 있
다4. 생비는 일반성 빈혈이 가 세포에 많음에도 불구
하고 결합생장아는 1:3 으로 여아에 많은 것으로
알려져 있다5.

1100년 영국에서 결합생장아가 처음으로 보고
되었으며, 이들은 둔부가 결합된 상태로 34세까지
생존하였다6. 지금까지 가장 널리 알려진 결합생장
아는 1811년 Siam에서 태어난 Eng와 Chang으로
상복부가 붙은 상태로 63세까지 생존하였으며, 이
들은 세커스 단장인 Barnum에 의해 세계에 널리
알려지게 되었고, 이들 이후 결합생장아가 Siam 쌍
둥이로도 불리우게 되었다7.

결합생장아는 생식학적으로 동일한 염색체 구성
을 갖는 동성의 monozygotic monochorionic twin
으로 태어 2주째에 embryo의 불완전한 분열과 관
련이 있다고 알려져 있다8,9. 결합생장아는 공유장기
의 종류에 따라 흉결합체(thoracopagus), 제대결
합체(omphalopagus), 좌굴결합체(pygopagus), 천
골결합체(ischiopagus), 두개결합체(craniiopagus)
로 분류되며, 그 분류상 결합체가 40%로 가장
많으며, 제대결합체가 33%, 천골결합체가 19%의
빈도를 보이고 있다(표 1)10.

Omphalopagus는 xiphopagus라고도 불리우며,
심한 혈액의 결합없이 상복부가 결합된 형태로 세
로 일꽃을 마주 대하고 있으며, 주로 간이나 소화기
관을 공유하고, 채식이나 제대발달 등을 동반하기도
한다11. 저지에 따라서는 일부 심낭만 공유하여도
결합체로 분류하는 경우도 있으나12, 심장이 공유하
지 않고 일부 심낭만 공유한 경우는 흉결합체로 보
기 어렵고, 이 환아들 경우에 있어서는 분리과정
에서 심낭의 심장이 개방되여 있었는데, 이것은 원
래 심낭의 심장은 공유하고 있었던 것인지 분리과정
에서 손상을 입은 것인지의 구별이 어려웠다.

대체적으로 분리의 적정기간은 9-12 개월로 보고
있으며, 분리가 1년 이상 지연될 경우는 각각의 목
<table>
<thead>
<tr>
<th>Type</th>
<th>Incidence (%)</th>
<th>Common organs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracopagus</td>
<td>40</td>
<td>Heart, liver, gastrointestinal tract</td>
</tr>
<tr>
<td>Omphalopagus</td>
<td>33</td>
<td>Liver, gastrointestinal tract</td>
</tr>
<tr>
<td>Pygopagus</td>
<td>19</td>
<td>Spine, genitourinary system, anorectum</td>
</tr>
<tr>
<td>Ischiopagus</td>
<td>6</td>
<td>Pelvis, genitourinary system, liver, gastrointestinal tract</td>
</tr>
<tr>
<td>Craniopagus</td>
<td>2</td>
<td>Brain</td>
</tr>
</tbody>
</table>

Table 2. Classification and Incidence of Conjoined Twins

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- Filler RM: Conjoined twins and their separation. Semin perinatol 10:82-91, 1986