Traumatic Atlanto-Occipital Dislocation—A Comprehensive Analysis of All Case Series Found in the Spinal Trauma Literature

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Traumatic Atlanto-Occipital Dislocation—A Comprehensive Analysis of All Case Series Found in the Spinal Trauma Literature

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ABSTRACT

Background: Traumatic atlanto-occipital dislocation (TAOD) is one of the most devastating traumatic injuries, generally associated with immediate death after high-energy trauma. The aim of this study was to perform a systematic literature review of all cases series of TAOD and present the current state of this entity.

Methods: A systematic literature review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. Only case series with at least 5 cases were included in the analysis. We focused on survival rates, diagnostic methods, delays in diagnosis, outcomes, and cases successfully treated nonoperatively.

Results: A total of 17 articles were included (16 retrospective and 1 prospective study) with 341 patients. Six studies included pediatric patients only. The mean Glasgow Coma Scale at admission was ≤8 in all studies. Many different diagnostic criteria were used, but none of them had high accuracy. The overall mortality rate was 34.8%, but the studies’ designs were heterogeneous (some included only survivors). A high rate of concomitant traumatic brain injury was documented in some studies. We found it interesting that some patients were treated with cervical immobilization (37/341; 10.8%), which was generally used in less unstable injuries; however, the majority of patients were managed with an occipito-cervical fusion (193/341; 56.5%).

Conclusions: TAOD is a devastating traumatic injury, with a high mortality rate. An MRI may be recommended when there are subtle findings of TAOD and a normal computed tomography scan, such as subarachnoid hemorrhage in the posterior fossa, upper cervical injuries, or consistent neurological findings. Further studies are necessary to identify patients with mild MRI findings and TAOD that may be managed nonoperatively.

Cervical Spine

Keywords: atlanto-occipital dislocation, occipito-cervical dislocation, craniocervical dislocation, traumatic, injury

INTRODUCTION

Traumatic atlanto-occipital dislocation (TAOD), also known as occipital-cervical dislocation, craniocervical dislocation, craniocervical dissociation, or internal decapitation, was first reported in 1908 by Blackwood in regard to a patient with complete neurological deficit with a postmortem analysis reporting an occiput dislocated forward on the spinal column.1

TAOD is one of the most severe and devastating traumatic injuries, generally occurring after high-energy traumas; it constitutes up to 31% of immediate deaths in motor vehicle fatalities according to autopsy studies.2.3.4 The death rate is so high that, according to a literature review5 published in 2005, only 135 survivors were reported from 1966–2001.

Historically, many radiological measurements based on simple plain x-rays were used to diagnosis TAOD, but they were clearly insufficient and inadequate to evaluate properly the craniocervical junction (CCJ).6 Currently, computed tomography (CT) scan is the exam of choice for initial screening of trauma patients, but even after a CT scan there are some anecdotal cases of delayed diagnosis of TAOD and late neurological deterioration.7–9 In addition, management of less-evident forms of TAOD, such as those with only MRI findings and normal or subtle findings on CT scan, require further elucidation because instability is not clear.10,11 Finally, clinical presentation may vary.
from normal neurological exam and no spinal cord injury to complete paralysis and respiratory failure and cardiac arrest. This can lead to a wide range of presentations that may make the diagnosis difficult, potentially confounded by the fact that most of these patients may also have severe traumatic brain injury (TBI).12

In this context, we performed a comprehensive systematic literature review of clinical studies discussing the characteristics, diagnosis, and management of patients with TAOD.

METHODS

A systematic literature review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines.13 Only case series with at least 5 cases were included and analyzed, without age limit. We focused on diagnostic methods, delays in diagnosis, the survival rates, neurological outcomes, and cases successfully treated without surgery.

Search Strategy

Due to the multiple terms used to refer to TAOD, we performed multiple searches in the PubMed database with different strategies. All existing studies up until April 29, 2020, were queried and reviewed. A total of 4 different searches were performed including cross-referenced articles.

The first search was performed using the following key words: “atlanto AND occipital dislocation AND traumatic,” obtaining 122 studies, with 13 selected. The second search was performed using the following key words: “cranio AND cervical AND dislocation,” obtaining 42 studies with 1 selected. The third search was performed using the key words “occipito AND cervical AND dislocation,” obtaining 91 studies, with 3 selected. Finally, a fourth search was performed using the key words “cranio-cervical AND dissociation,” with an additional 4 studies obtained. Six studies were duplicates and thus excluded, for a total of 15 eligible studies. Additional cross-referencing provided 2 more clinical studies, for a total of 17 fully included. A flowchart detailing our search selection is presented in Figure 1.

Methodological Quality Evaluation: Potential risk for bias was assessed according to the Methodological Index for Non-Randomized Studies (MINORS) instrument (whereby fewer than 12 points suggest low-quality studies).14 All the studies were then analyzed for internal validity integrity and graded for level of evidence in accordance to the Oxford Centre for Evidence-Based Medicine.15

Eligibility Criteria: Clinical case series, retrospective or prospective, with a minimum 5 cases of TAOD were included. Studies should be written in English language; small case series or narrative and systematic reviews were excluded.

Data Collection Process

One of us (A.F.J.) independently extracted data from the included studies using a piloted data extraction form.

Data Extraction

The following data were extracted from the included studies, when described: number of patients, study design, patients’ characteristics (age, Glasgow Coma Scale [GCS], sex), diagnosis and radiological findings, interventions performed, outcomes and follow-up, results, and survival rates.

RESULTS

Of the 17 included articles, 16 were retrospective case series (1 was a retrospective case series from a prospective trauma database—level 4 evidence) and only 1 was a prospective study (level 2 evidence).8–10,12,16–28 In Table 1 we summarized all the data extracted from the 17 articles fully included. All studies except 2 had a MINORS score of fewer than 12 points, suggesting high risk for bias (see Table 2).17,24

EPIDEMIOLOGY

A total of 341 patients were included in the studies. Of those, 119 patients died (just before admission or during the follow-up). The overall mortality rate was 34.8%, but it may not reflect the reality, because some studies excluded nonsurvivors or nontreated patients. The survival rate in the oldest included study, published by Cohen et al,16 reported 100% mortality at 1 year. Six of the 17 articles included only children and adolescents, with a total of 78 patients (78/341 = 22.8% of the entire patient sample), and there was a mortality rate of 55.1% (43/78 patients during the studies’ follow-up).9,12,16,17,20,27
With the exception of 2 studies (both on pediatric patients), all had a majority of patients who were men.12,27

Mean GCS at admission, when reported, was 8 points or fewer in all studies, compatible with comatose patients (6.6 points in the Astur et al9 study, 8 points in the Filiberto et al26 study, 7.4 points in the Hosalkar et al20 study). Individual GCS at admission ranged from 3 points (some of the patients were in cardiac arrest) to 15 points in some studies.

Delayed diagnosis was reported in 22 cases, generally with neurological deterioration.8,9,23 The study of Reis et al23 reported that delays in diagnosis decreased from 1996–2002 to 2003–2008, suggesting that advances in radiological imaging may increase the diagnosis rate at admission. Of note, the criteria for delayed diagnosis were not clear in the studies. We inferred that early diagnoses were those made immediately after the initial radiological evaluation.

RADIOLOGICAL FINDINGS

We noted a tremendous heterogeneity in the diagnostic criteria used in the included studies. In 2 of the studies, radiological methods to diagnose were not detailed: Hosalkar et al20 reported that the diagnosis was made using the Power ratio, the Kaufman method, and the Harris method. Filiberto et al26 reported that diagnosis was made by an attending neuroradiologist using CT or MRI scans.
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<tr>
<th>Author and Study Design</th>
<th>N and Neurological Status at Admission</th>
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<th>Results/Outcome</th>
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<tbody>
<tr>
<td>Cohen et al,16 1991 Retrospective case series</td>
<td>5 patients Mean of 5 y 3 boys and 2 girls 1 died on admission, 4 in cardiac arrest</td>
<td>AOD clear in x-rays in 3 patients, inconclusive in 2 (basion or opisthion not defined on the plain films)—CT diagnosis 5 had prevertebral hematoma</td>
<td>CT indicated SAH in 3, edema in 1</td>
<td>4 tetraplegia 5 multiple injuries 1 died on admission, 2 survived for a few hours, 2 for months (5 and 8 months)</td>
</tr>
<tr>
<td>Bulus et al, 1993 Retrospective case series</td>
<td>11 patients Mean age of 5 y (range, 1.5–12.5 y) 6 presented with cardiac arrest (GCS of 3) and 5 were alert and responsive, 3 of them neurologically intact</td>
<td>Normal BDI in 10 lateral cervical spine radiographs was 8.3 ± 4.2 mm and all 11 had &gt;1.4 cm (mean = 1.7 mm) Power ratio &gt;1 in only 6 patients 9 had prevertebral swelling 5 had C1-C2 subluxation 2 had C1 or C2 fractures 9 patients had CT scan: 6 had SAH, 4 cerebral edema, 3 cerebral contusion, 1 intraventricular hemorrhage, and 2 skull fractures</td>
<td>5 survivors were immobilized with a halo (4) or soft collar (1) and 1 required an OCF due to persistent instability</td>
<td>6/11 died (within 5 d after admission) 5/11 survivors with minimal or no neurologic sequel cranial palsies solved when involved cranial nerves VI and VII, but not XII</td>
</tr>
<tr>
<td>Ahuja et al,18 1994 Retrospective case series</td>
<td>6 patients all &lt; 30 y (11–28 y, mean = 25.6 y) 5 men and 1 woman (2 children) 2 had asystole and 4 agonal respirations</td>
<td>x-rays with retropharyngeal swelling in all patients, Power ratio was &gt;1 in 5/6 3 patients had late diagnosis (1–3 days after injury) 3 patients had unilateral occipital condyle fractures 3 had cerebral edema, basal ganglia hemorrhage, and 2 skull fractures</td>
<td>Halo in 3 patients, Philadelphia collar in 2, and Crutchfield tongs in 1 3 had subsequent OCF and 2 who had halo were treated with halo for 3 mo</td>
<td>patient who had Crutchfield tongs developed quadriplegia and respiratory failure due to overdistraction and died of sepsis 1 mo later 5 survivors had cranial nerve palsies or mild hemiparesis</td>
</tr>
<tr>
<td>Przybylski et al,19 1996 Retrospective case series</td>
<td>7 patients Median 18 y (range, 5–60 y), 1 was neurologically intact and 2 had hemiplegia</td>
<td>axial CT scan: 6/7 with SAH around the brain stem and upper cervical spinal cord, with blood in the 4th ventricle too; none had SAH 3/7 had AOD diagnosed with x-rays Power ratio was &gt;1 in 3/7 patients</td>
<td>4 treated with halo and OCF and 1 had a OCF 5 mo after halo vest use</td>
<td>5 young (range, 2–15 y) were alive at a median 4-year follow-up 2 oldest died within 3 days of injury</td>
</tr>
<tr>
<td>Kenter et al,12 2001 Retrospective case series</td>
<td>5 patients 3 girls (60%) and 2 boys (40%) mean age = 8 y (range, 7–12 y)</td>
<td>Wholey and Power measurements were performed in the initial plain radiographs 3 were diagnosed based on x-rays; 1 was diagnosed using CT and 1 MRI (both had SCI) none of the 5 had fractures and only 2 had retropharyngeal swelling</td>
<td>3 survivors were initially immobilized with a halo and then had an OCF</td>
<td>4/5 had TBI—all 4 intubated 2/5 died (24 h and 4 d, respectively) 4/5 had spinal cord injury 3 survivors: 2 intact, 1 remained quadriplegic 2 nondiagnosed AOD using initial cervical lateral x-rays but were retrospectively diagnosed AOD when the x-rays were reviewed</td>
</tr>
<tr>
<td>Hosalkar et al,20 2005 Retrospective case series</td>
<td>16 patients Mean age = 7.6 y (range, 1.3–16.3 y) Mean GCS at admission was 7.4 points (3–15)</td>
<td>authors used the lateral radiographs of the cervical spine for screening, using the Power ratio, the Kaufman method, and the Harris method.</td>
<td>8/16 of survivors were immobilized with orthosis/halo; 3 of them died later from TBI, 4 of the remaining 5 had an OCF (Occ-C2) and 1 a Minerva cast</td>
<td>11/16 intubation in the field, 2/16 in the emergency department (ED), and 3 were not intubated 8/16 declared dead on arrival in the ED 15/16 received steroids in the ED of the 5 survivors, 1 was neurologically normal, 3 had spastic hemiparesis and were functional, and 1 was quadriplegic and ventilator-dependent</td>
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<tr>
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<td><strong>Horn et al,</strong> 2007</td>
<td>33 patients, mean age = 21.1 y (range, 1–55 y)</td>
<td>All had CT scan (2.5 mm) and 23/33 had an MRI (those with grossly abnormal CT findings, an MRI was not performed) normal/ near normal CT and abnormal MRI findings: diagnosis of OAD when there is abnormal signal in the occipital condyle-atlantal lateral mass joint capsule or posterior ligamentous structures (posterior atlanto-occipital membrane and ligamentum nuchae)</td>
<td>BDI had 77% sensitivity and BAI-BDI had 73% sensitivity</td>
<td>18/33 (55%) had atlantoaxial dissociation</td>
</tr>
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<td><strong>Cooper et al,</strong> 2010</td>
<td>69 patients with 47 diagnosed postmortem, 22 diagnosed in hospital</td>
<td>19/22 survivors had abnormal BAI or BDI on radiographs, whereas 2 had abnormal BAI or BDI on cervical CT images</td>
<td>7 survivors had OCF</td>
<td>7/22 survivors (higher GCS, more likely normotensive, and none had cervical cord injury)</td>
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<td><strong>Kim et al,</strong> 2012</td>
<td>5 patients, Mean age = 34.6 y (range, 24–50 y)</td>
<td>3 had early diagnosis and 2 late (both deteriorated neurologically) all cases: prevertebral soft tissue swelling at C2 cervical x-ray: mean = 17.88 mm (range, 15.18–20.17 mm; nl, 4 mm)</td>
<td>5 (100%) had OCF</td>
<td>Delayed diagnosis: neurological deterioration all 5 discharged from hospital</td>
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<td>Radcliff et al,10 2012</td>
<td>18 patients 18/4519 = 0.3% age range, 18–83 y 15 men and 3 women</td>
<td>displacement: considered when measures was outside 3 SD CI of normal no patient had Oc-C1 dislocation without atlantoaxial displacement 13/18 had displacement outside the normal range, 6 with displacement of both Oc-C1 and C12 joints, with 5 with displacement of C12 joints and 2 with abnormal BDI only. 12 had abnormal BDI, 7 without major articular displacement 17 had MRI, 11 with injured cruciate ligament, 4 with indeterminate injury and 2 with an intact ligament 5 patients with Oc-C1 articular displacement had injury to the Oc-C1 capsule</td>
<td>all had OCF</td>
<td>3 patients with Oc-C1 dislocations had complete cord injury and 3 patients were intact 5 patients with atlantoaxial dislocations and patients without displacement or ligamentous injury were intact 4 AIS A, 3 AIS D, and 11 intact.</td>
</tr>
<tr>
<td>Astur et al,9 2013</td>
<td>14 patients Mean age = 5.2 y (range, 1–10 y) Mean GCS at admission 6.6 (3–15) 7 boys (50%) and 7 girls (50%)</td>
<td>OCF from 1 to 14 days (mean 3.8 days) OCF all (Oc-C1 in 3, Oc-C2 in 5, Oc-C3 in 3, and Oc-C4 in 3 patients) 6 patients had dural tears 4/5 used halo for initial treatment 2 had surgery 3/5 died early (on days 2, 7, and 11)</td>
<td>12 had associated injuries (11 had TBI) 4 patients (28.5%) had hydrocephalus after OCD; all had a ventriculoperitoneal shunt 6 mo: 8 patients had neurological impairment, 6 patients required a tracheotomy and mean hospitalization was 24.9 d (range, 6–75 d) 1 had incomplete tetraplegia without needing ventilation 1 able to walk 3/5 (60%) had associated OCF</td>
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<td>Mueller et al,24 2013</td>
<td>5 patients 3/5 died due to the severity of the injury mean age was 37.8 y (range, 15–52 y) 0.2% of the trauma GCS range from 3 (3 who died) to 11 and 15 points not specified: injury diagnosed by an attending neuroradiologist using CT or MRI Power ratio &gt;1 in 1/5</td>
<td>38/52 (73%) had stabilization 17 had OCF, 10 external orthosis, and 11 both 3 (18%) of 17 deaths had some stabilization 35 survivors had some stabilization versus 18% of the nonsurvivors (P &lt; .001)</td>
<td>34 (97%) of survivors were neurologically intact and 1 discharged with neurological deficits GCS at the admission: only predictor of death (OR 0.7; 95% CI 0.552-0.877); mean of 4 points (nonsurvivors) vs 10 (survivors); P &lt; .001 deaths: 94% due to TBI</td>
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<td>Menon et al,25 2017</td>
<td>5 patients 3 men and 2 women age ranged from 23–33 y (no children)—mean = 27.8 y</td>
<td>5 OCF (occiput-C3/4)</td>
<td>4 were independently mobile and 1 wheelchair bound (GCS 3 at admission) Mean of 2.5 y of follow-up 1/5 had occipital condyle fracture surgery day 1 to 4 33% mortality (17 non survivors versus 35 survivors)</td>
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<td>Filiberto et al,26 2018</td>
<td>52 patients Mean age of 44 (15 to 85 years) Mean admission GCS of 8 35 men (67%) and 17 women (33%)</td>
<td>5 OCF (occiput-C3/4)</td>
<td>4 were independently mobile and 1 wheelchair bound (GCS 3 at admission) Mean of 2.5 y of follow-up 1/5 had occipital condyle fracture surgery day 1 to 4 33% mortality (17 non survivors versus 35 survivors) 34 (97%) of survivors were neurologically intact and 1 discharged with neurological deficits GCS at the admission: only predictor of death (OR 0.7; 95% CI 0.552-0.877); mean of 4 points (nonsurvivors) vs 10 (survivors); P &lt; .001 deaths: 94% due to TBI</td>
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### Table 1. Continued.

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<tr>
<td>Hazboun et al.(^{27}) 2019 Retrospective case series</td>
<td>27 patients 10 died (37%) 11 boys and 16 girls Median age was 5 y (60 mo; range, 2–109 mo)</td>
<td>all had cervical spine radiographs at admission; and BDI, BAI, Power ratio and CCI were assessed. 17 survivors: none of them diagnosed using x-rays; 10 diagnosed by CT (41%) and 10 (59%) by MRI—7 of them not diagnosed on initial plain x-rays or CT imaging but had persistent neck pain; 9/10 MRI had prevertebral soft tissue swelling and apical ligament disruption, with tears in the tectorial membrane 14/17 survivors had some evidence of brain injury 10 nonsurvivors: diagnosed by plain radiograph (5) and CT scan (the remaining 5) with SAH along the clivus and hypoxia-induced encephalopathy</td>
<td>all 17 survivors had an OCF</td>
<td>Survivors—median = 13.4 mo of follow-up; 4 quadriplegic prior to surgery; 8 discharged intact, 8 discharged with some degree of spinal cord injury, and 5 with deficits secondary to TBI Median time to mortality was 1.5 d (1–7 d) survivors had significantly less cardiopulmonary resuscitation ((P &lt; .01)), lower injury severity scores ((P &lt; .01)), higher GCS scores ((P &lt; .001)) and shorter transport times ((P &lt; .01)). 9 (53%) had no disability at follow-up evaluation BDI was significantly greater in the nonsurvivors</td>
</tr>
<tr>
<td>Chang et al.(^{28}) 2020 Retrospective case series</td>
<td>15 patients 13 men and 2 women Mean age = 50.3 y (range, 21–93 y) 1 was quadriplegic, 2 comatose and the others were neurologically intact</td>
<td>AOD was diagnosed by Wackenheim line and BDI on CT, MRI or x-rays 11 posterior type AOD and 4 vertical type AOD 4 had brain injuries (subdural, posterior fossa, ventricular, and intracerebral hemorrhage).</td>
<td>5 OCF, 4 received a halo vest, and 4 were transferred using a Philadelphia brace</td>
<td>8 (72.7%) had C1 fractures, 4 (36.4%) had C1 lateral mass fractures, 2 (18.2%) had transverse atlantal ligament injuries. 2 (50%) patients of those with vertical AOD had C12 vertical subluxation; 2 patients had type II dens fractures; 1 C6-C7 fracture dislocation and other C3 fracture. 1 died 2 d after admission</td>
</tr>
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</table>

Abbreviations: AIS, American Spinal Injury Association Impairment Scale; AOD, atlanto-occipital dislocation; BAI, basion-axial interval (basion from the posterior cortex of the axis)—horizontal distance between the basion and the posterior cortex of the axis; BDI, basion-dental interval—distance between the basion and tip of the dens; CCI, occipital condyle to C1 interval; DBI (or BDI), distance between the apex of dens and basion ≤ 12 mm; GCS, Glasgow Coma Score; MVA, motor vehicle accident; n1, normal; Oc–C1, the occipital joints (condyles) and C1 joint; OCD, occipito-cervical dislocation; OCF, occipito-cervical fusion; OR, odds ratio; SAH, subarachnoid hemorrhage; SCI, spinal cord injury; TBI, traumatic brain injury.
We summarized some of the most-used radiological criteria, as well as their descriptions, in Table 3.

Cohen et al,\textsuperscript{16} in the oldest published study (1991) included in our review, reported that 2 of 5 (40\%) patients had inconclusive x-rays due to limitation in the visualization of the basion and opisthion. It is interesting that 5 of 5 (100\%) patients had a prevertebral hematoma. This study included only children. The same rate of retropharyngeal swelling was reported by Ahuja et al\textsuperscript{18}; all 6 children had retropharyngeal swelling at the x-rays. On the other hand, only 2 of 5 (40\%) children included in the Kenter et al\textsuperscript{12} study had retropharyngeal swelling, with contradictory incidences.

Bulas et al\textsuperscript{17} also reported a high rate of prevertebral hematoma (9/11; 81.8\%) in their series. All 11 patients had a basion-dens interval (BDI) \(\geq 1.4\) cm (the normal BDI, according to the authors and based on 110 normal lateral cervical radiographs used as controls, was \(8.3 \pm 4.2\) mm). We found it interesting that the Power ratio was abnormal (>1) in 6 of 11 patients (54.5\%); comparatively, the Power ratio was abnormal in 3 of 5 (60\%) of the patients in the Mueller et al\textsuperscript{24} study, 1 of 5 patients (20\%) in the Menon et al\textsuperscript{25} study, and 3 of 5 (60\%) patients in the Kim et al\textsuperscript{8} study.

Cooper et al\textsuperscript{22} reported that a BDI \(\geq 16\) mm was associated with nonsurvivors (\(P = .004\)), and 21 of 22 patients had an abnormal BDI or basion-axis interval (BAI)—the exception was 1 patient with C0-C1 widening despite a normal BDI and BAI.\textsuperscript{22} The same comparison was performed in the study of Hazboun et al,\textsuperscript{27} who reported a higher BDI in nonsurviving pediatric patients than in those who survived.\textsuperscript{27}

A high rate of upper cervical spine injuries were associated with TAOD in some articles, similarly to TBI, such as occipital condyle fractures, C1-C2 subluxation, C1 and C2 fractures, and transverse atlantal ligament injuries, among others.\textsuperscript{17,18,28} Bulas et al\textsuperscript{17} reported that 50\% of their patients (3/6) had unilateral occipital condyle fractures.

The following diagnostic criteria were used or cited in the included studies: BDI and BAI, Power ratio, Wholey method, prevertebral swelling, Kaufman method, Harris method, MRI findings of ligamentous injuries, displacement outside 3 SDs of normal measurements of the C0-C1 joints, the Wackenhein clivus baseline, and finally, the Pang rule.
Hazboun et al. reported that some patients (7/17; 41.1%) were not diagnosed after x-rays and CT scan but due to persistent neck pain underwent an MRI, and this identified TAOD. This was the only study emphasizing a high failure rate on diagnosing TAOD with x-rays and CT requiring MRI.

TRAUMATIC BRAIN INJURIES

Many studies reported a high rate of TBI associated with TAOD. Cohen et al. reported 3 of 5 patients (60%) with subarachnoid hemorrhage (SAH) and 1 patient with brain edema evident in the brain CT scan. In their series of 6 patients, Ahuja et al. documented 3 patients with cerebral edema, 1 with intracerebral hemorrhage, and 4 with SAH. In the Przybylski et al. study, 6 of 7 patients had SAH (85.7%). Eleven of 15 (73.3%) had TBI in the Astur et al. study of pediatric patients. It interested us that in their study, 4 pediatric patients (28.5%) with TAOD required a ventriculoperitoneal shunt for hydrocephalus.

In the Hazboun et al. study, CT scan findings were more evident and severe, even with hypoxia induced by encephalopathy, in nonsurvivors (10/10 had brain CT scan changes).

Finally, in the most recent included study by Chang et al. in 2020 with 15 patients, 4 had intracranial injuries.

NEUROLOGICAL STATUS

Of the 222 survivors (65.1%), about 46.8% (104) were neurologically intact or almost intact (mild deficits); 11.2% (25) had severe deficits, tetraplegia, and/or ventilator dependence, and in 41.8% (93) the details of the neurological status in the follow-up were not obtained. Some studies reported neurological deficits due to TBI alone or associated with spinal cord injury, which were not always possible to characterize in detail.

We found it curious that many studies reported cardiac arrest in the emergency room or just after admission.

MANAGEMENT

When treatment was mentioned, an occipitocervical fusion (OCF) was performed in 193 patients, with or without a postoperative cervical orthosis or halo. If we included all 341 patients, 56.5% were able to receive an OCF. On the other hand, 37 patients (10.8%; 37/341) were treated with a cervical orthosis, halo vest, or Crutchfield tongs and did not have surgery. In a series of 33 patients of Horn et al. the rationale for nonoperative management was clarified: patients with abnormal findings on MRI with no or minimal injuries evident on CT scans. Five patients were treated according to these criteria and 4 had follow-up; none of them required further surgery.

A total of 341 patients were included in the studies. Of these, 119 patients died (just before admission or during the follow-up). The overall mortality rate was 34.8%, but it may not reflect the reality, because some studies excluded nonsurvivors or nontreated patients. The survival rate in the oldest included study, published by Cohen et al. reported 100% mortality in 1 year. Of the 17 articles, 6 included only children and adolescents, with a total of 78 patients (78/341 = 22.8% of the entire patient

Table 3. Summary of the mainly used diagnosis criteria for traumatic atlanto-occipital dislocation (TAOD) in the included studies as well as their description.

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<td>BAI</td>
<td>Basion-axial interval—distance of the basion from the posterior cortex of the axis</td>
</tr>
<tr>
<td>BDI (Wholey method)</td>
<td>Basion-dens interval (also described as DBI: dens-basion interval)—distance from the tip of the dens to the basion—normal range &lt;10 mm</td>
</tr>
<tr>
<td>Power ratio</td>
<td>Ratio between the distance from the basion and the posterior spinolaminar line of the atlas/the distance from the opisthion and the anterior arch of the atlas (&gt;1—anterior TAOD should be suspected)</td>
</tr>
<tr>
<td>Prevertebral swelling</td>
<td>Generally &lt;10 mm in the upper cervical spine</td>
</tr>
<tr>
<td>Kaufman method</td>
<td>The actual distance between the occipital condyle and the condylar facet of C1 (the normal range width of the atlanto-occipital junction is never &gt;5 mm)</td>
</tr>
<tr>
<td>Harris method</td>
<td>BAI in children was more reliable due to the lack of ossification of the dens before 12 y of age. Normal BAI extends from 12 mm anterior to 4 mm to the posterior axial line</td>
</tr>
<tr>
<td>MRI findings of ligamentous injury</td>
<td>Direct visualization of ligamentous injuries or lack of articular congruence in the MRI</td>
</tr>
<tr>
<td>Pang rule: the condyle-C1 interval</td>
<td>CCI is the measurement of the interval between the occipital condyle and C1 at 4 equidistant points on the joint surface in sagittal and coronal reconstructions of CT (a total of 8 measurements for each patient). The true CCI consisted in the mean of the 8 readings for both sides in coronal and sagittal images; with a cutoff of 4 mm (the CCI is highly symmetrical in children).</td>
</tr>
<tr>
<td>Traynelis classification</td>
<td>Type I—anterior dislocation of the cranium, type II—vertical dislocation/distraction, and type III—posterior dislocation</td>
</tr>
</tbody>
</table>
DISCUSSION

We performed one of the most comprehensive systematic literature reviews of TAOD. Despite its limitations from including different study designs, such as some studies evaluating only survivors and others including all patients with a diagnosis of TAOD, it provides some interesting insights about this rare but devastating entity. It is widely known that TAOD should be treated surgically in the vast majority of patients as soon as possible. The mortality rate of 34.8%, reaching up to 68.7% in an isolated study, is quite high. TAOD generally affects men, who present as comatose and with concomitant TBI at the emergency department, despite a small number of patients who may be neurologically intact. In children, it seems to affect equally boys and girls.

Zivot et al reported 26 of 85 (31%) atlanto-occipital dislocations as the immediate cause of death in fatal motor vehicle–pedestrian accidents. The instantaneous death may be due to brainstem injury, resulting in vascular and neurogenic shock, respiratory failure, and immediate death, potentiated by concomitant injuries, such as TBI or multisystem trauma. In the retrospective study of a prospectively collected database included in our review, Radcliff et al reported that the incidence of TAOD among all spinal traumas in their institution was 18 patients of a total of 4519 spinal trauma cases—representing 0.3% of the entire sample.

Children are more susceptible to TAOD than adults, and, in our review, they also had a higher mortality rate. There are potential reasons for a higher incidence of TAOD in this age group: a higher relationship between head size/spine proportion (the head size/spine proportion is substantially larger in children than in adults), the immature composition of the spine (more mobile osseous and musculoligamentous composition), and the plane of the atlanto-occipital joint, which is practically horizontal with small condyles. All these reasons together may put children at a higher risk of TAOD. We also believe that the lower bone density facilitates ligamentous injuries instead of bone fractures, explaining a higher rate of ligamentous injury in children than in adults.

Theodore et al reported in a systematic review that nearly 20% of the patients with acute TAOD had a normal neurological exam at admission. Although the final outcome was not detailed in all included articles, overall they reported that 46.8% of patients were neurologically intact or presented with mild deficits at the last follow-up or at hospital discharge, compared with 11.2% patients who were severely disabled (in 41.8% of the cases details of the clinical outcome were not provided). For this reason, early detection and treatment is paramount for patients who survive initial trauma and are able to receive prehospital care. In many studies, cranial nerve palsy were also documented, most with clinical improvement over time. The most common cranial nerve injuries associated with TAOD described were to the VI, VII, and XII.

The potential explanations for these injuries are sudden distraction or even injury to the nerve nuclei itself in the brainstem.

Radiological Methods to Diagnose TAOD

In our review, there was a tremendous heterogeneity in the radiological methods used to diagnose TAOD. This was also consistent with the pertinent literature about challenges in the radiological diagnosis of TAOD. There were many proposed plain radiographic methods to diagnose TAOD. The most used included (1) a displacement of more than 10 mm between the basion and tip of the dens—the BDI; (2) the ratio of the basion to the midpoint of the anterior cortex of the posterior arch of C1 distance divided by the ratio of the opisthion to the midpoint of the posterior cortex of the anterior arch of C1, which should be greater than 1 to diagnose—Power ratio; (3) a distance from the posterior mandible and anterior atlas of more than 13 mm or a distance from the posterior mandible and dens of more than 20 mm; (4) Failure of a line from the basion to the axis spinolaminar junction to intersect C2 or a line from the opisthion to the posterior inferior corner of the body of the axis to intersect C1—the X-line method; (5) a displacement of more than 12 mm or less than 4 mm between the basion and posterior C2 line (BAI) or a displacement of more than 12 mm from the basion to the dens (BDI). These measurements included studies published in 1958 and 1979, with the newest one published in 1994, which did not consider the quality of modern CT scan image and reconstructions. The limitations to proper bone and joints evaluation, as well as the low sensitivity (about 0.505) and specificity of x-rays in diagnosing TAOD.
Traumatic Atlanto-Occipital Dislocation

scan with reformatted images is the initial line of imaging for trauma patients and radiographs should be used only if CT is unavailable or unaffordable. In the same position statement protocol, they suggested that MRI should be indicated in patients with neurological involvement and advanced cervical degenerative changes to evaluate epidural-space compromise, disco-ligamentous injury, and also soft tissue injuries. Figures 2–4 illustrate the most important radiological measurements for diagnosing TAOD.

In 2013, a guideline of the American Association of Neurological Surgeons concluded that initial lateral cervical radiographs may miss the diagnosis of TAOD, which was also observed in some cases published in our review, resulting in later neurological deficit. By that time, they suggested that a high index of suspicion should be maintained in patients with prevertebral soft tissue swelling on “normal” lateral x-rays or in patients with cranio-cervical SAH on axial CT images. They also stated that there was a high level of evidence to use the condyle-C1 interval (CCI) in pediatric patients to diagnose TAOD. This index was published by Pang et al with a sensitivity and specificity of 100% compared with standard tests on plain films that had sensitivities varying from 25%–50% and specificities from 10%–60% in diagnosing TAOD.

Figure 2. Sagittal midline computed tomography (CT) scan measuring the distance from the basion to the tip of the dens—the basion-dens interval. Normal values from adults’ CT scans are <8.5 mm in 95% of the cases.

Figure 3. (A) Sagittal and (B) coronal computed tomography (CT) scan reconstructions—4 equidistant points were marked on the articulating surfaces of the occiput-C1 joint that encompassed the broadest joint surface. The joint interval at each point was recorded and the mean of these 8 measurements (from both sides and from sagittal and coronal CT scans) are the true condyle-C1 interval (CCI).
The CCI is the measurement of the interval between the occipital condyle and C1 at 4 equidistant points on the joint surface in sagittal and coronal CT reconstructions (a total of 8 measurements for each patient). The true CCI consisted in the mean of the 8 readings for both sides in coronal and sagittal images; with a cutoff of 4 mm (the CCI is highly symmetrical in children).

More recently, Gire et al.\(^3\) reported a radiological study designed to evaluate diagnostic measurements, with 10 cases of TAOD and 10 cases of non-TAOD. They evaluated the sensitivity, specificity, and reliability of 8 radiographic criteria applied to CT to diagnosis TAOD. They defined 2 new measurements: the revised CCI (measured at the point of greatest separation between the occipital condyle and C1 lateral mass, perpendicular from the surface of the lateral mass to the surface of the occipital condyle—TAOD was diagnosed when the left and/or right CCI was >2.5 mm) and the condylar sum (which consisted of the sum of the left and right CCI and TAOD was diagnosed when \( \geq 5 \) mm). The revised CCI and the condylar sum were highly sensitive (100%), specific (92% for condylar sum and 84% for CCI), and reliable (intraclass correlation coefficients of 0.88 and 0.93 for CCI and the condylar sum, respectively) for detecting TAOD when using a CT scan.\(^3\) The same was concluded by Dahdaleh et al.\(^2\) who performed another diagnostic study using CT scans comparing the BDI, the BAI, Lee X-lines, Power ratio, CCI, and revised CCI. They concluded that the revised CCI method was the most simple, sensitive, and reliable method. The high sensitivity and specificity of accessing directly the occipital-C1 joint is probably because the measurements directly assessed TAOD, whereas the other measurements were indirect evaluations. Of note, these “gold standard” CT scan measurements were not applied in any clinical series included in our systematic review, thus requiring further studies.

Martinez-del-Campo et al.\(^4\) in 2016, tried to identify the normal CCI in healthy adults and compare these values with the CCI in adults with a known diagnosis of TAOD to establish more sensitive and specific cutoff values for diagnosing TAOD. They evaluated 59 adults without TAOD and 22 with TAOD, measuring thin-slice CT scans to assess the CCI, condylar sum, the Wholey and Harris intervals, the Power and Sun ratios, and the Wackenheim line and Lee X-lines. Patients without TAOD included 30 men (50.8%) and 29 women (49.2%) with a mean age of 42 ± 16 years (range, 19–87 years), and the group with TAOD comprised 10 men (45.5%) and 12 women (54.5%) with a mean age of 38.2 ± 9.7 years (range, 20–56 years). The mean CCI obtained for non-TAOD patients was 0.89 ± 0.12 mm, with the largest one measuring 1.4 mm. The mean condylar sum was 1.8 ± 0.2 mm, with the largest condylar sum value of 2.2 mm. In

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**Figure 4.** Sagittal CT scan (A) of a normal and (B) of a patient with a traumatic atlanto-occipital dislocation (TAOD). The red line in B represents the revised condyle-C1 interval (CCI). The diagnosis of TAOD is made when distance at the point of great separation of the C1 lateral mass from the occipital condyle is greater than 2.5 mm.\(^4\)
patients with TAOD, the mean CCI was 3.35 ± 0.18
mm (range, 1.5–6.4 mm). The shortest single CCI
measurement in the TAOD patients was 1.1 mm.
Martinez-del-Campo et al40 reported that the mean
condylar sum for all 22 TAOD patients was 6.7 ±
2.7 mm and the shortest condylar sum was 3.0 mm.
After these measurements, the authors proposed
new cutoff values for AOD: 1.5 mm for the CCI and
3.0 mm for the condylar sum in adults (compared
with 4 mm in children for the CCI), with a
sensitivity of 1 and false-negative rate of 0. The
sensitivities of other measurements were 0.55, 0.46,
0.27, 0.23, 0.41, and 0.41 for Power, Wholey, Harris,
Sun, Wackenheim, and Lee criteria, respectively.

The role of MRI in TAOD is still debated. The
evaluation of ligamentous injury in MRI has
supplanted any other image modality, but in the
setting of a normal or near-normal CT, these
findings may not be enough to indicate an OCF.
Horn et al21 tried to evaluate the role of short TI
inversion recovery sequences in determining cervical
spine instability after trauma in 314 patients. They
correlated soft tissue edema with x-rays and CT
findings and concluded that when CT and x-rays did
not demonstrate evident instability, MRI did not
provide additional information. In the Hazboun et
al27 study, however, the authors reported that 7
patients (of 17 survivors, a total of 41.1%) had
normal initial cervical x-rays or CT spine scans but,
due to persistent neck pain, underwent an MRI with
the diagnosis of a TAOD requiring an OCF. This
was also proposed by a radiological study of
Corcoran et al41 that reported that CCI was not
effective to diagnosis some cases of TAOD. The
sensitivity used by Pang et al33 (4 mm) was low and
the specificity of Gire et al39 for their revised CCI
(2.5 mm) was also low for less severe forms of
TAOD but clinically significant. For these reasons,
they stated the importance of MRI for complete
evaluation of these patients.

Abouelleil et al42 reported an interesting case of
TAOD wherein the CCI was normal, but there was
severe TAOD diagnosed with an increased BDI (19
mm) and atlantoaxial vertical subluxation. TAOD
was clearly confirmed using an MRI, with evident
apical ligamentous disruption at the craniovertebral
junction (CVJ) and C1-C2 distraction. They42
proposed that in the presence of C1-C2 dislocation,
the CCI interval may not diagnose TAOD, and
other measurements such as the BDI and MRI
should be considered. It is important to note that
the BDI cutoff for diagnosing TAOD on the basis of
a sagittal CT scan is <8.5 mm in 95 of 100 normal
adults, compared with 12 mm on plain radiographs.43
We also believe that some patients with
bilateral condyle avulsion may have normal CCI or
revised CCI and severe forms of TAOD.

Treatment Considerations—Nonoperative
Management

Horn et al21 described 5 cases treated non-
operatively based on abnormal findings on MRI
but with mild or no changes evident on the CT
scans. They proposed that external immobilization
would be an option when there is a normal CT with
mild findings on MRI, such as moderate to severe
injury to posterior interspinous and atlanto-occipital
ligaments and mild injury to the C0-C1 joints.
They stated that the dilemma: to overtreat some
patients with normal CT scan measurements but
C0-C1 joint disruptions on MRI or not to treat
them, which may lead to catastrophic neurological
injuries or even death. To solve this dilemma, they
proposed a classification based on 2 grades: grade 1,
characterized by normal CT scan findings but
moderate MRI findings (high posterior ligaments
or occipitoatlantal signal) that may be treated
nonoperatively; and grade 2, minimum of 1
abnormal CT scan finding of established diagnostic
criteria and grossly abnormal findings in MRI, such
as in the occipitoatlantal joints, tectorial membrane,
aral ligaments, or cruciate ligaments, which may be
surgically fixed. It seems reasonable to not treat all
patients who have only MRI findings but also to fix
some patients who are at risk of a severe cata-
strophic injury. Although the authors did not
discuss this in their study, we believe that patients
with grade 1 but with spinal cord injury should be
aggressively managed with OCF, similar to spinal
cord injuries at any other level.

Kaplan et al44 published a literature review of
nonoperative management of craniocervical liga-
mentous distraction injuries. They presented a case
of a 27-year-old man who had severe ligamentous
injury at C0-C1 and C1-C2 treated successfully with
a halo for 6 months due to his poor clinical
condition, with reduction and evident stability on
dynamic x-rays and minimal symptoms at 41
months after his injury. They attributed their
successful outcome to the fact that the patient
probably had only incomplete injury to the main
ligaments of the upper cervical spine. The outcomes
of nonoperative management are not clear, and the risks potentially outweigh the benefits. Child et al\textsuperscript{45} studied the craniocervical ligament’s ability to prevent craniocervical distraction in a cadaveric biomechanical model using a fluoroscopic traction test and successive sectioning of the primary ligamentous complexes. Six specimens from occiput-C3 had the following ligaments sectioned to simulate a TAOD: alar, tectorial membrane, and occiput-C1 capsules. Traction was applied using 0–20 lb with fluoroscopy, and digital images analysis quantified the displacement from C0-C1, craniocervical excursion, and under what load a 2-mm displacement could be reproducible. It is interesting that minimal dislocation was reported with sectioning 2 of 3 ligaments (<2 mm). They reported that specimens had a firm restraint to dissociation until all 3 ligaments were cut—in an “all-or-none” restraint to instability characteristic. For these reasons, all 3 ligamentous complexes (ie, alar ligaments, tectorial membrane, and atlanto-occipital capsules) may be important to maintain stability in anatomical models and may explain why some patients may survive neurologically intact.

The commonly cited classification for TAOD published by Traynelis et al\textsuperscript{34} divided injuries in 3 types: type I (anterior dislocation of the occipital condyles in relationship with the atlas lateral masses), type II (longitudinal dislocation of the condyles), and type III (posterior dislocation of the condyles in relationship with the atlas lateral masses).\textsuperscript{34} This classification was criticized by Horn et al\textsuperscript{21} due to the potential hypermobility of these injuries, which would allow all 3 categories to be displayed in the same patient. There is also a possibility that such instability could be well positioned, and this may preclude evidencing the displacement in x-rays or CT scan.\textsuperscript{23}

A significant number of studies reported other spinal injuries associated with TAOD, such as upper spinal fractures. In their series, Radcliff et al\textsuperscript{10} emphasized the high prevalence of atlantoaxial dislocations, which may or may not be associated with TAOD. In 5 cases, C1-C2 dislocations were not associated with TAOD, suggesting that separated structures stabilized the occipitoatlantal and atlantoaxial ligaments. Treatment for these associated injuries should be taken into account in a tailored approach.

Treatment Considerations—Operative Management

As already stated, the great majority of TAOD should be surgically treated as soon as possible. Although preoperative cranial traction may be used to realign the CCJ, up to 10% neurological deterioration has been reported in the literature, which may require extreme cautions and close clinical and radiological follow-up if traction is intended.\textsuperscript{30}

When operative treatment is chosen for TAOD, modern occipito-cervical fixation techniques are based on screw-rod hardware, with a high rate of fusion and immediate occipito-cervical stabilization.\textsuperscript{36} Without associated subaxial injuries, an occipital C2 screw fixation is generally enough to restore CCJ stability. The C2 fixation can be achieved using different techniques, such as laminar, pars, pedicle, or transarticular screws, considering the patient’s specific anatomy and the surgeon’s experience.\textsuperscript{47}

Traumatic Brain Injuries

Because the CVJ is stable due to strong ligamentous structures, a high-energy impact is generally necessary to cause TAOD. For this reason, it is commonly associated with facial trauma and TBI, which may occult the spinal cord injury once patients present as comatose to the emergency department, which may also contribute to the delay in diagnosis in some patients.\textsuperscript{12,20} In addition, some patients may die due to TBI and not from the TAOD.\textsuperscript{22} Different types of intracranial pathology were documented in our review, such as cerebral edema, brain contusion, intraventricular hemorrhage, subdural hematoma, and the most common, SAH, especially in the posterior fossa, which may lead to a high suspicion of AOD.\textsuperscript{30} SAH in front of the pons, for instance, could lead to a high suspicion of CCJ pathology, especially because CT and MRI are made in a supine position and blood can flow up to the brain. On the other hand, cortical SAH is common in TBI and may not lead to a specific suspicion of TAOD.

Unfortunately, this review has several limitations: First, only English literature was reviewed, the quality of the included studies was not high, and we only used the PubMed database for screening. Finally, studies performed in the 1980s and 1990s may have different results from more recent studies,
due to advances in neuroradiological exams with better anatomical details and image quality as well as improvement in surgical techniques to stabilize the occipito-cervical junction when compared with old fixation devices. We did not perform an individual analysis according to the year of the study. In spite of these limitations, we believe that our review provides a comprehensive view of TAOD that may be useful for designing future prospective studies.

CONCLUSIONS

TAOD is a devastating traumatic injury, with a high mortality rate and patients presenting as comatose in the majority of cases. There is no pathognomonic radiological diagnostic criteria, especially when CT findings are not so evident, although the CCI is promising, especially when associated with the BDI. MRI may be recommended when there are subtle findings of TAOD and a normal CT, such as SAH in the posterior fossa, upper cervical injuries, or consistent neurological findings. Surgical treatment is recommended in the majority of cases. Further studies are necessary to identify patients with mild MRI findings and TAOD who may be managed nonoperatively.

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