



Review Article

Surgical management of dropped head syndrome: A systematic review

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ABSTRACT

Background: Dropped head syndrome (DHS) is uncommon and involves severe weakness of neck-extensor muscles resulting in a progressive reducible cervical kyphosis. The first-line management consists of medical treatment targeted at diagnosing underlying pathologies. However, the surgical management of DHS has not been well studied.

Methods: Here, we systematically reviewed the PubMed and Cochrane databases for DHS using the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. All relevant articles up to March 31, 2022, were analyzed. The patient had to be ≥ 18 years with DHS and had to have undergone surgery with outcomes data available. Outcomes measurements included neurological status, rate of failure (RF), horizontal gaze, and complications.

Results: A total of 22 articles selected for this study identified 54 patients who averaged 68.9 years of age. Cervical arthrodesis without thoracic extension was performed in seven patients with a RF of 71%. Cervicothoracic arthrodesis was performed in 46 patients with an RF of 13%. The most chosen upper level of fusion was C2 in 63% of cases, and the occiput was included only in 13% of patients. All patients neurologically stabilized or improved, while 75% of undergoing anterior procedures exhibited postoperative dysphagia and/or airway-related complications.

Conclusion: The early surgery for patients with DHS who demonstrate neurological compromise or progressive deformity is safe and effective and leads to excellent outcomes.

Keywords: Cervical kyphosis, Cervical spine, Dropped head syndrome, Isolated neck extensor myopathy, Spine surgery

INTRODUCTION

Dropped head syndrome (DHS) is rare and involves severe weakness of the cervical extensor muscles resulting in a progressive reducible severe kyphosis.^[1-3,7,9] Patients symptomatically present with; neck pain, gait disturbances, myelopathys, difficulty eating, inability to lift the head, and impaired horizontal gaze.^[3] DHS can occur in association with, generalized neuromuscular

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diseases, isolated neck extensor myopathy,^[2,7,9] parkinsonism, hyper/hypothyroidism, spinal cord injury, myelopathy, some malignancies, secondary to radiotherapy, and/or medications such as mitogen-activated protein kinase inhibitors. Notably, DHS is passively correctable in its early stages and patients may favorably respond to nonoperative treatments.^[3] Thus, comprehensive workups to are critical before proceeding with surgical interventions.^[3,10] Our aim was to analyze the current literature on the surgical management of DHS and its clinical and radiographic outcomes.

MATERIALS AND METHODS

A comprehensive systematic review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines.^[9] A National Library of Medicine (through PubMed) and Cochrane Library database research were performed by two independent reviewers (M.J.C. and J.M.O.). The search terms included: “*dropped head syndrome*,” “*isolated neck extensor myopathy*,” “*cervical deformity*,” “*cervical kyphosis*,” and “*chin on chest*” with AND and OR combinations. All relevant articles published in English up to March 31, 2022, with full text were included in the study. We utilized multiple inclusion and exclusion criteria [Table 1].

Methodological quality assessment

Methodological quality assessment was performed using a tool specifically designed by Murad *et al.*^[8] to assess case reports and case series. This scale is based on eight items that can be categorized into four domains: selection, ascertainment, causality, and reporting. The maximum number for each item was: 1 for selection, 2 for ascertainment, 4 for causality, and 1 for reporting. All the included articles scored five or more points.

Table 1: Inclusion and exclusion criteria.

Inclusion Criteria	Exclusion Criteria
Age ≥ 18	Pediatric population
Diagnosis of DHS who underwent surgical treatment	Patients without surgical treatment
Prospective or retrospective case series, cohorts, or case reports	Technical notes, literature reviews, conference abstracts, and cadaveric studies
At least one treatment outcome reported (e.g., neurological improvement, cervical alignment, horizontal gaze, complications, treatment failure, and surgical revision)	Patients with fixed cervical deformity, ossification of the posterior longitudinal ligament, or ankylosing spondylitis.
Documented of follow-up.	
DHS: Dropped head syndrome	

RESULTS

Inclusion of 22 articles

We finally chose 22 articles for analysis [Figure 1]. Of those, 15 were case reports, and seven were retrospective case series. Fifty-four patients (37 females and 17 males) with DHS syndrome, who were surgically treated, were identified and the mean age at the time of surgery for the cohort was 68.9 years old (range 38–88 years) with a mean of follow-up of 16.1 months (range 3–72 months).

Symptoms

The most common symptoms were inability to lift the head and impaired horizontal gaze with and without gait disturbance (100% of patients). Neurological deficits were present in 11 patients (20.37%), and nine (16.66%) patients presented with difficulty swallowing. Most of the patients also reported preoperative neck or back pain (57.41%).

The presentation of symptoms was gradual in most patients ranging between 1 month and 20 years of preoperative symptoms. Of the articles that reported prior treatments, conservative nonsurgical treatments failed in 25 patients.

Surgical strategies

Surgical strategies were divided into two major groups: cervical arthrodesis alone without thoracic extension (CA) and cervicothoracic arthrodesis (CT). In each strategy, patients were further treated with either posterior fusion alone (PF), anterior release with PF, or anterior fusion with PF (combined approach). Forty-six patients (85% of the analyzed cohort) underwent CT surgery (22 with combined fusion, five anterior releases with PF, and 19 with PF alone) and seven CA surgeries.

CT findings

The most common upper instrumented cervical level was C2 (63%); the occiput was included only in 13% of the patients. The most common lower instrumented vertebra in the thoracic spine was T3 (26%) followed by T4 (21%). Only three of the 46 patients who had CT fusion had an extension of the construction to the lower thoracic levels. With regard to the patients undergoing CA (cervical arthrodesis alone without thoracic extension), surgery included the combined approach in four patients, C3–C6 PF in 1, C2–C6 laminoplasty, and C4–C6 anterior corpectomy and reconstruction in one patient [Table 2].

C7 pedicle subtraction osteotomies were performed on five patients and posterior vertebral column resection in one patient.

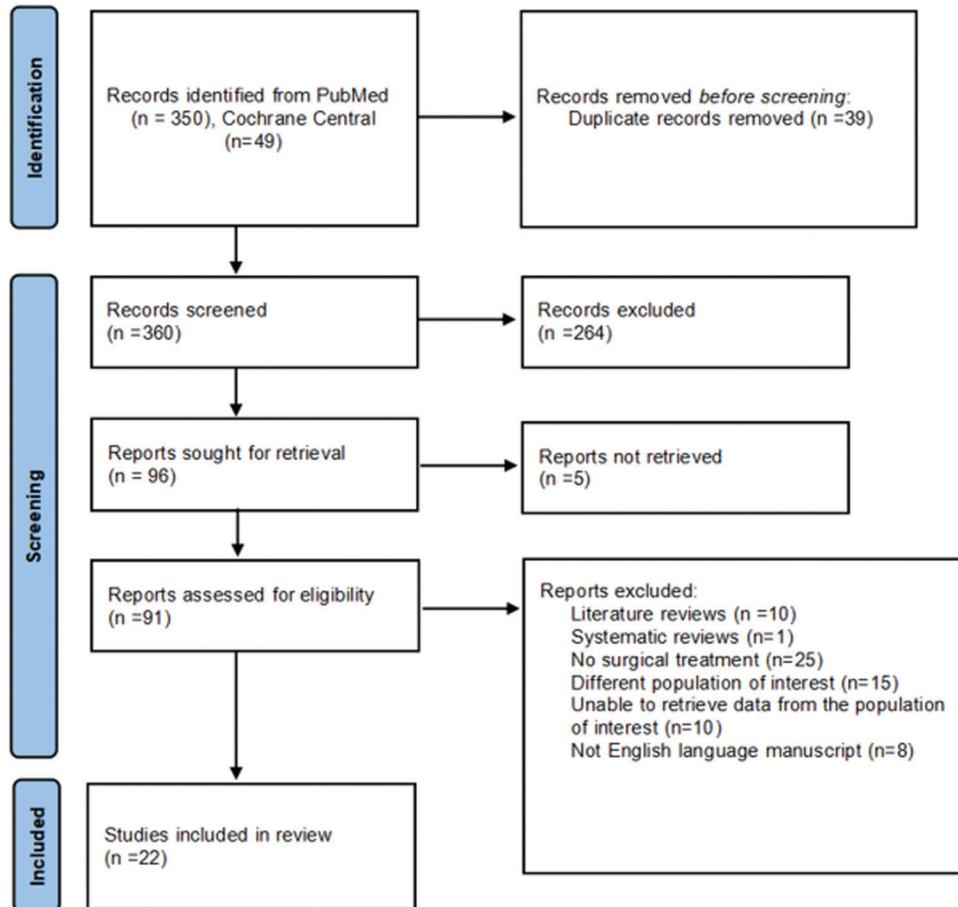


Figure 1: Flow diagram showing study selection process following PRISMA guidelines.

Postoperative neurological outcomes

Neurological examinations improved or remained intact for all patients independent of the approach chosen. Most patients (91.3%) with CT surgery showed improvement in horizontal gaze. Surgical failure was defined as the need for revision surgery for kyphosis progression, impaired horizontal gaze, distal junctional kyphosis, and/or pseudarthrosis.

Surgical failure

The rate of failure for patients with CA was 71%. Surgical failure was observed in six patients (13%) with CT, four with a combined approach, and two with PF. Both patients with failure after PF were treated with wires.

Postoperative surgical and medical complications

Multiple postoperative surgical and medical complications were observed in these 54 patients with DHS. Surgical site infection was observed in one patient (1.8%), one patient expressed limited neck extension after surgery. Twelve medical complications were observed in 11 patients and

included five with dysphagia, four with postoperative pneumonia, two with respiratory distress, and one with sepsis. Most medical complications were dysphagia or airway-related complications. About 75% of dysphagia or airway-related complications were observed in patients with combined approaches for anterior fusion or anterior release.

Radiographic outcomes

The main sagittal parameter analyzed in this systematic review was the cervical (C2–C7) angle. Pre- and postoperative C2–C7 angle was reported in 41 patients [Table 3]. Most patients showed a significant improvement in cervical kyphosis after surgery. Only one patient showed a similar pre- and postoperative C2–C7 angle related to surgical failure.

Sagittal vertical axis (SVA) was defined as negative SVA if was < 0 cm and positive SVA if was $SVA \geq 0$ cm or P-SVA.^[5,6] SVA correction was described in 27 patients [Table 4]. Surgical failure was observed in only one patient with N-SVA and in five patients with P-SVA.^[6] Two patients with preoperative P-SVA, without surgical failure, complained of back pain after surgery.

Table 2: Studies including patients with DHS treated with surgery.

S. No.	Author	N	Follow-up (months)	Surgery	Etiology	Indication	Horizontal Gaze	Surgical Failure	Complications	Sagittal Parameters
1.	Rust, 2011	1	24	C3–C6 combined (CA)	SCI	Failed PT	Not improved	Yes	Progressive Kyphosis -Revision surgery	Not specified
2.	Koda, 2015	2	Not specified	CT PF Combined CT	Postoperative INEM	Not specified	Improved	No	None	C2–C7 angle, LL, T1 slope
3.	Petheram, 2008	1	18	CT PF	INEM	No specified	Improved	No	Limited neck extension	Not specified
4.	Kudo, 2020	15	12	Combined CT (13) CA combined (2)	No specified	No specified	Improved (14) Not improved (1)	Yes (4 CT, 2 CA)	DJK (3) Need for an extension to lumbar levels (3) Respiratory distress (1) Dysphagia (1) SSI (1)	C2–C7 angle, LL, T1 slope, SVA
5.	Nakanishi, 2007	1	12	Occipito-CT PF	INEM	Failed therapy with steroids	Improved	No	None	Not specified
6.	Verla, 2021	7	Not specified	Combined CT (1) Combined Occipito-CT (1) CT PF (4) Occipito-CT PF (1)	RDT	Failed PT and/or bracing	Improved	No	Dysphagia (2) Sepsis (1) Pneumonia (2)	C2–C7 angle, SVA
7.	Amin, 2003	1	3	PF CT	Epilepsy	Not specified	Improved	No	None	Not specified
8.	Marcon, 2018	1	3	Combined CT	INEM	Failed PT and bracing	Improved	No	None	Not specified
9.	Odate, 2018	1	15	C4–C6 corpectomy (CA)	Postoperative	Failed PT and/or bracing	Not improved	Yes	Progressive Kyphosis -Revision surgery	Not specified
10.	Bronson, 2018	1	13	Combined CT	INEM	Not specified	Improved	No	None	C2–C7 angle,
11.	Gerling, 2009	9	72	Combined CT (1) CT PF (5) CT AR and PF (3)	RDT INEM	Failed PT and/or collar Failed steroids or IVIG	Improved (5) Not improved (2)	Yes (2 CT)	Progressive Kyphosis -Revision surgery (2) Pneumonia (2) Dysphagia (2) Pulmonary Edema (1)	C2–C7 angle
12.	Caruso, 2014	1	19	CT PF	RDT	Not specified	Improved	No	None	C2–C7 angle
13.	Kawaguchi, 2004	1	48	C2–C6 laminoplasty (CA)	Myelopathy	Not specified	Improved	No	None	Not specified
14.	Rahimizadeh, 2016	1	18	C2–C6 combined	INEM	Failed PT and bracing	Not Improved	Yes	Progressive Kyphosis -Revision surgery (2)	Not specified

(Contd...)

Table 2: (Continued).

S. No.	Author	N	Follow-up (months)	Surgery	Etiology	Indication	Horizontal Gaze	Surgical Failure	Complications	Sagittal Parameters
15.	Martin, 2011	1	8	Occipito-CT PF	INEM	Failed PT and bracing	Improved	No	None	Not specified
16.	Stoker, 2012	1	12	CT PF	After Radiofrequency	Not specified	Improved	No	None	Not specified
17.	Farshad, 2021	1	12	Occitopexy	RDT	Not specified	Improved	No	None	Not specified
18.	Takahashi, 2011	1	12	CA combined	Myelopathy	Failed medical and bracing treatment	Improved	No	None	C2–C7 angle,
19.	Kusakabe, 2020	1	Not specified	Combined CT	Arthritis Rheumatoid	Failed PT and bracing Failed steroids	Improved	No	None	C2–C7 angle, LL, T1 slope, SVA
20.	Katiriji, 2005	1	14	CT PF	Myopathy	Failed PT and/or bracing	Improved	No	None	Not specified
21.	Pereira, 2010	1	Not specified	Occipito-CT PF	Parkinson	Not specified	Improved	No	None	Not specified
22.	Hashimoto, 2018	4	Not specified	Combined CT (1) Combined Occipito-CT (1) CT AR and PF (2)	No specified	Not specified	Improved	No	None	C2–C7 angle, LL, T1 slope, SVA

DHS: Dropped head syndrome, AR: anterior release, CA: cervical-only approach without extension to the thoracic spine, CT: cervicothoracic fusion, DJK: distal junction kyphosis, INEM: isolated neck extensor myopathy, LL: lumbar lordosis, N: number of patients in the study, PF: posterior fusion, PT: physical therapy, RDT: radiotherapy, SSI: surgical site infection, SVA: sagittal vertical axis

DISCUSSION

Etiology of DHS

DHS is characterized by either marked neck extensor weakness,^[9] or dystonia primarily of the deep cervical flexor muscles with kyphosis that can be passively reduced. Cervical paraspinal muscles contribute to the neck stability and mobility, and the radicular innervation of this muscle is segmental, but typically begins cranially at the C3 root.

Treatment of DHS

The treatment of DHS relies on a full etiological work-up, and initial treatment is typically nonsurgical.^[2,3,7] A previous systematic review showed an optimal positive response of 87.5% with medical treatment and immunosuppression. Surgical indications for DHS should be carefully evaluated.^[2] Because this disorder affects the elderly population, a growing prevalence is expected given the continued aging of the population.^[3,7] We found 54 patients with DHS who were surgically treated.

The surgical treatment of DHS aims to restore the horizontal gaze, correct sagittal alignment, and decompress neural elements. In this study, we found a 71% failure rate for patients fused only within the cervical spine (CA patients) to treat DHS.

Successful clinical and radiological outcomes of DHS with CA were only found in two patients (29%) in this systematic review. Of those, Kawaguchi *et al.* described one patient with cervical stenosis and myelopathy with DHS who greatly improved after cervical laminoplasty. The other patients described by Takahashi *et al.* underwent C4–C6 corpectomy and C3–C7 posterior fusion.

N-SVA in DHS has been described as a spinopelvic compensation to the cervical deformity and is associated with a better response to surgery.^[5,6] Conversely, patients with P-SVA tend to have larger preoperative T1 slopes, larger C2–C7 SVA, and less lumbar lordosis.^[6] Higher rates of failure in P-SVA patients have been shown by Kudo *et al.*^[6] and some authors recommend more extensive fixation or additional surgeries in these patients to the concomitant thoracic kyphosis.^[6]

Table 3: Studies reporting pre and postoperative C2–C7 angle.

Author	Approach	N	Preoperative C2–C7 angle (°)	Postoperative C2–C7 angle (°)	Degree of correction (°)
Kudo, 2020	Combined CT (13) CA combined (2)	15	Mean–47.3	Mean 15.3	Mean 62.6
Verla, 2021	Combined CT (1) Combined Occipito-CT (1) CT PF (4) Occipito-CT PF (1)	7	Mean–21.5	Mean–0.03	Mean 21.62
Gerling, 2009	Combined CT (1) CT PF (5) CT AR and PF (3)	9	Mean–69	Mean–28	Mean 41
Koda, 2015	CT PF	1	–35.8	17.7	53.5
Koda, 2015	Combined CT	1	–50	22.2	72.2
Bronson, 2018	Combined CT	1	–35	10	45
Caruso, 2014	CT PF	1	7.9	38.3	30.4
Hashimoto, 2018	Combined CT (1) Combined Occipito-CT (1) CT AR and PF (2)	4	Mean–38.25	Mean 10	Mean 48
Takahashi, 2011	CA combined	1	–47	–20	27
Kusakabe, 2020	Combined CT	1	–76	3	79

AR: Anterior release, CA: cervical-only approach without extension to the thoracic spine, CT: cervicothoracic fusion, N: number of patients in the study, PF: posterior fusion

Table 4: Studies reporting pre- and postoperative SVA.

Author	Approach	N	Preoperative SVA (cm)	Postoperative SVA (cm)	Degree of correction (cm)
Kudo, 2020	Combined CT (13) CA combined (2)	15	Mean–0.7	Mean 2.21	Mean 2.95
Verla, 2021	Combined CT (1) Combined Occipito-CT (1) CT PF (4) Occipito-CT PF (1)	7	Mean 6.96	Mean 3.94	Mean –3.02
Hashimoto, 2018	Combined CT	1	–1.1	4	5.1
Hashimoto, 2018	CT AR and PF	1	–2	–0.3	1.7
Hashimoto, 2018	CT AR and PF	1	5	7	2
Hashimoto, 2018	Combined Occipito-CT	1	3	1.6	–1.4
Kusakabe, 2020	Combined CT	1	–6°	–0.5	5.5

AR: Anterior release, CA: cervical-only approach without extension to the thoracic spine, CT: cervicothoracic fusion, N: number of patients in the study, PF: posterior fusion, SVA: sagittal vertical axis

Extension of fusion to upper thoracic spine

The extension of the fusion to the upper thoracic segments between T1 and T5 may provide a more stable biomechanical support to the cervical construct and reduces the risk of failure.^[2] In this review cervical fusion to the upper thoracic spine, restored the horizontal gaze and maintained or improved neurological status in 91.3% and 100% of the patients, respectively.^[2,4] Moreover, the failure rate of a construct that crosses the cervicothoracic junction was 13% compared to 71%, as mentioned above.

DHS usually shows an enlarged C2–C7 angle, but usually maintained the C0–C2 angle. In this systematic review, C2 was the most chosen upper instrumented level, and the occiput was included only in 13% of the constructions. In addition, an extension of the fusion to the occiput significantly restricts head movements and can lead to significant rates of complications. A recent study showed a 44.5% rate of complications for DHS patients undergoing occipital-cervical fusion.

In this systematic review, combined approaches for deformity correction were slightly preferred over PF. This is likely as

combined approaches allow for better restoration of the lordosis. However, medical complications such as dysphagia and those related to the airway were observed most frequently in patients with anterior approaches for fusion or release. Thus, the use of a combined approach for DHS must be carefully weighed against potential early complications after surgery.

In general, rigid cervical deformities are associated with a greater number and grade of osteotomies than flexible deformities; however, surgical osteotomies may be necessary to correct later stages of the disease or severe cases of DHS.^[2] Because most of the studies did not report the use of posterior column osteotomies, the risk or advantage cannot be analyzed.

The amount of deformity correction wanted must be weighted with the risk of complications and expected functional outcomes when deciding between combined approaches, PF, and/or different grades of osteotomies.

Finally, surgical treatment is not the first treatment strategy for DHS, but the likelihood of needing surgery seems to increase when the cause is unknown thus highlighting the importance of a longitudinal follow-up for these patients.

CONCLUSION

The early surgery for patients with DHS who demonstrate neurological compromise or progressive deformity is safe, effective, and leads to excellent outcomes.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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