

## Case Report

# Natural History, Neuroradiological Workup, and Management Options of Chronic Atlantoaxial Rotatory Fixation Caused by Drug-Induced Cervical Dystonia

Yuki Ishikawa,<sup>1</sup> So Kato <sup>1</sup>, Mario Ganau <sup>2</sup>, Shima Hirai,<sup>1</sup> Yasushi Oshima,<sup>1</sup> and Sakae Tanaka<sup>1</sup>

<sup>1</sup>Department of Orthopaedic Surgery, The University of Tokyo, 113-8655, 7-3-1, Hongo, Bunkyo-ku, Tokyo, Japan

<sup>2</sup>Department of Neurosurgery, Oxford University Hospitals NHS Foundation Trust, Oxford, UK

Correspondence should be addressed to So Kato; sokato@g.ecc.u-tokyo.ac.jp

Received 29 October 2020; Accepted 18 February 2021; Published 3 March 2021

Academic Editor: Johannes Mayr

Copyright © 2021 Yuki Ishikawa et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Atlantoaxial rotatory fixation (AARF) resulting from drug-induced cervical dystonia (DICD) represents an extremely rare complication of antipsychotic treatment, requiring a comprehensive assessment of pharmacologic therapy and timely radiologic workup. We report a chronic case of Fielding type I, Pang type I AARF secondary to schizophrenia treatment in a 16-year-old girl, along with a review of the literature on the management challenges posed in this condition. In this scenario, torticollis may just represent the tip of the iceberg, and only an effective multidisciplinary approach increases the chances of satisfactory correction with closed reduction, hence avoiding the burden of more invasive treatment options.

## 1. Introduction

The rotatory subluxation of the atlantoaxial joint, also known as atlantoaxial rotatory fixation (AARF), is a frequent cause of torticollis in children [1], which was firstly reported by Corner in 1907 [2]. A thorough clinical description of this condition was provided by Wortzman and Dewar in 1968 [3]; nonetheless, the term AARF was introduced only by Fielding and Hawkins who also classified AARF into 4 types (Table 1) in 1977 [4]. Over the years, this condition has been described as atlantoaxial rotatory subluxation [3, 5, 6], dislocation [7], or displacement [8], making the pertinent literature fairly heterogeneous until the seminal work from Pang led to the introduction and diffusion of a more modern classification and management algorithm (Table 1) [9].

Patients with AARF present with neck pain, limited neck motion, and the so-called cock-robin posture, where the head is rotated to one side and on lateral flexion con-

tralaterally (Figure 1(a)); the most common causes are trauma and infection, rheumatoid arthritis, head and neck surgery, and congenital connective disorders [10, 11].

Rarely, cervical dystonia has been linked to AARF [12–14]. Although it is known that antipsychotics sometimes induce cervical dystonia [15, 16], there have been no reports of AARF caused by drug-induced cervical dystonia (DICD). We report such a rare presentation during schizophrenia treatment, enriched by a review of the literature.

## 2. Case Report

**2.1. Clinical History.** A 16-year-old girl with recent diagnosis of schizophrenia was prescribed treatment with antipsychotics. After few months, she started to have neck pain and torticollis and two years later was diagnosed with antipsychotic-related tardive DICD. An antiparkinsonian agent was started, and botulinum toxin was injected twice,

TABLE 1: AARF classifications.

Fielding and Hawkins AARF classification [4]	
Type 1	Rotatory fixation with no anterior displacement
Type 2	Rotatory fixation with anterior displacement of 3 to 5 millimeters
Type 3	Rotatory fixation with anterior displacement of more than 5 millimeters
Type 4	Rotatory fixation with posterior displacement
Pang AARF classification [9, 17]	
Type 1	AARF patients show essentially unaltered (“locked”) C1-C2 coupling regardless of corrective counterrotation
Type 2	AARF patients show reduction of the C1-C2 separation angle with forced correction, but C1 cannot be made to cross C2
Type 3	AARF patients show C1-C2 crossover but only when the head is cranked far to the opposite side

yielding minimal improvement. After a year of unsuccessful treatment, she was hospitalized for further treatment and was referred to our department for evaluation of the torticollis.

**2.2. Examination and Diagnostic Workup.** Due to the uncontrolled schizophrenia, she had delusions, hallucinations, and disorganized thinking and behaviors. Although communication was very difficult, the patient was identified to be suffering from persistent neck pain and difficulties with eating, swallowing, and gaze. She was fixed in a typical cock-robin posture; the neck was twisted to the right and flexed to the left (Figure 1(a)). She had no neurological deficits.

Computed tomography (CT) revealed Fielding type I and chronic Pang type I AARF [4, 17] (Figures 1(b)–1(h)). The atlas was rotated 60 degrees to the right in the axial view (Figures 1(b) and 1(c)). The lateral inclination of the atlas was 25 degrees in the coronal view (Figure 1(d)). The atlanto-dental interval (ADI) was 2 mm in the sagittal view (Figure 1(e)). In 3D reconstruction images, the C1-C2 joint was locked, although C2 facet deformity was not confirmed (Figures 1(f)–1(h)).

**2.3. Treatment Course.** Considering her mental condition, continuous traction, halo vest, and surgical intervention were not deemed appropriate in the first instance, due to safety concerns. She was mentally unstable and had a risk of taking unpredictable movements; hence, the consensus from multidisciplinary team (MDT) discussion was to perform only closed manual reduction followed by immobilization with the Philadelphia collar. Closed manual reduction was conducted under general anesthesia and fluoroscopy (Arcadis Orbic 3D, Siemens Healthineers) in the operation room. Neck alignment appeared normalized in the neutral position (Figure 2(a)), and reposition of the atlas was confirmed with fluoroscopy (Figures 2(b) and 2(c)). The patient was placed in a Philadelphia collar fixation immediately.

**2.4. Outcome.** Unfortunately, few days later, torticollis recurred despite good compliance with the Philadelphia collar. The atlas was rotated 45 degrees to the right from the neutral position (Figure 3). The strategy adopted included an immediate attempt to achieve reduction again under general anesthesia; nonetheless, this also led to an iatrogenic fracture of C2 lateral mass (Figure 4(a)) and atlantoaxial rotatory subluxation remained (Figures 4(b) and 4(c)). Family discussion was held, and surgical option was minimized the chances of recurrence or pseudoarthrosis was abandoned given the high perioperative risks. The final decision was therefore to continue conservative treatment with the Philadelphia collar and carry out a close clinical and radiological monitoring. Bony union was obtained 2 months later (Figure 5), and during follow-up, the patient demonstrated a consistent improvement of neck pain, range of neck motion, and activities of daily living (ADL); therefore, no further spinal treatment was needed. Five months after the fracture, range of neck motion had improved to 60 degrees, which was considered to be a good result, given that the C1-C2 joint was already completely “locked” before treatments.

### 3. Discussion

There have been no previous reports of AARF with drug-induced cervical dystonia. Tardive dystonia is known as one of the side effects of antipsychotics, and the most frequent type is cervical dystonia [16, 18]. The first choice of treatments of cervical dystonia is oral medications such as trihexyphenidyl, benzodiazepines, or tetrabenazine. Other choices are botulinum injection and deep brain stimulation [19–22]. Although the mechanisms underlying AARF in patients with cervical dystonia are not clearly understood, it is assumed that the contraction of the sternocleidomastoid muscle causes rotatory subluxation of the atlas and the axis.

Pang’s classification, which further groups patients into acute, subacute, and chronic, is particularly useful to guide the treatment strategy. Whereas acute (<1 month) and subacute (>1- $<3$  months) AARF can usually be treated by nonoperative treatment such as closed manipulation or cervical traction followed by bracing [10, 23], the treatment strategy for chronic AARF (lasting >3 months) is controversial. Halter traction and skull caliper traction are recommended as the initial treatment in the algorithm by Pang and Li [23]. Ishii et al. reported that closed manipulation followed by halo fixation is an effective treatment. Surgical treatments are recommended for cases with C1-2 bony union, irreducible or recurrent subluxation, and Fielding type 4 [10]. Treatment options of AARF are shown in Table 2 [10, 24–28].

The complexity of treating patients with uncontrolled schizophrenia relates to their tendency toward unpredictable movements and higher rates of postoperative complications [29, 30]. Any invasive treatments, such as traction and surgery, are therefore not recommended. When treating this challenging category of patients, it is important to ensure a comprehensive diagnostic workup and start

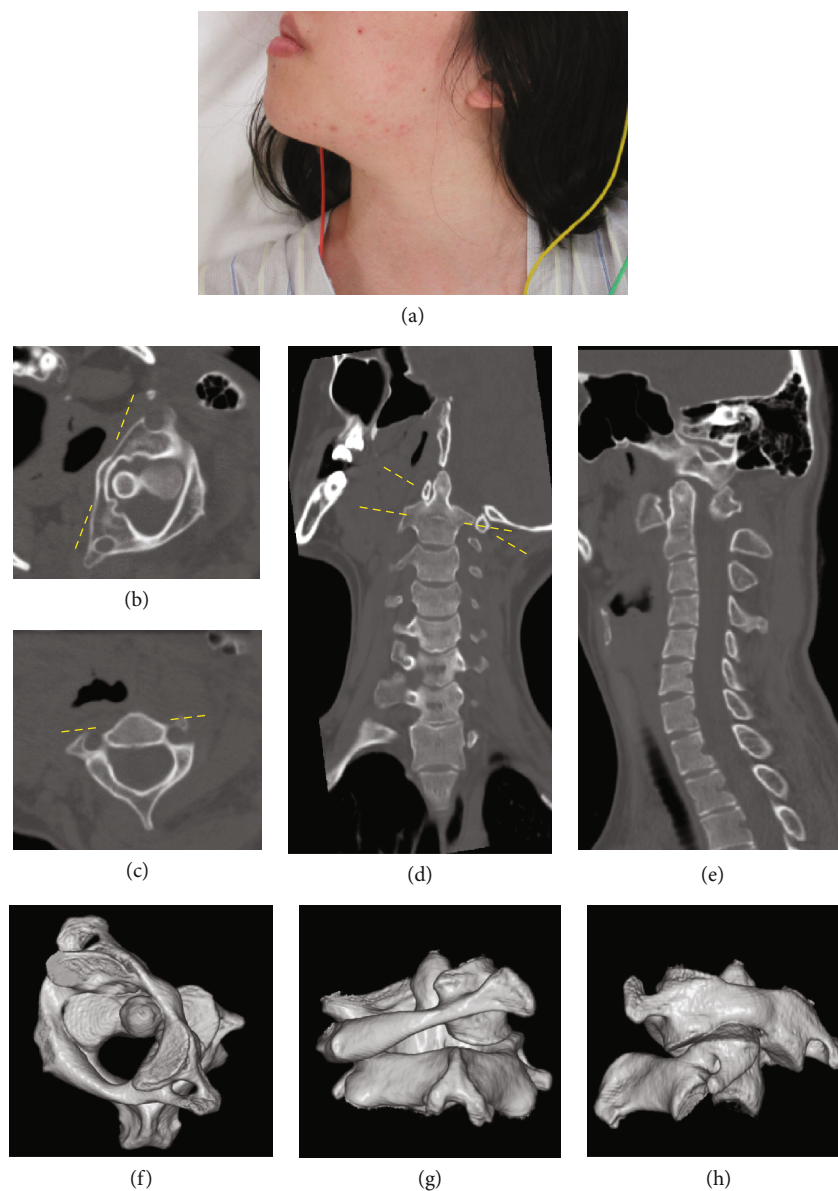


FIGURE 1: Patient position before reduction (a) and CT of the cervical spine on admission (b-h). (b) Axial view of C1, (c) axial view of C2, (d) coronal view, (e) sagittal view, and (f-h) three-dimensional reconstruction. The C1-C2 rotatory angle was 60 degrees.

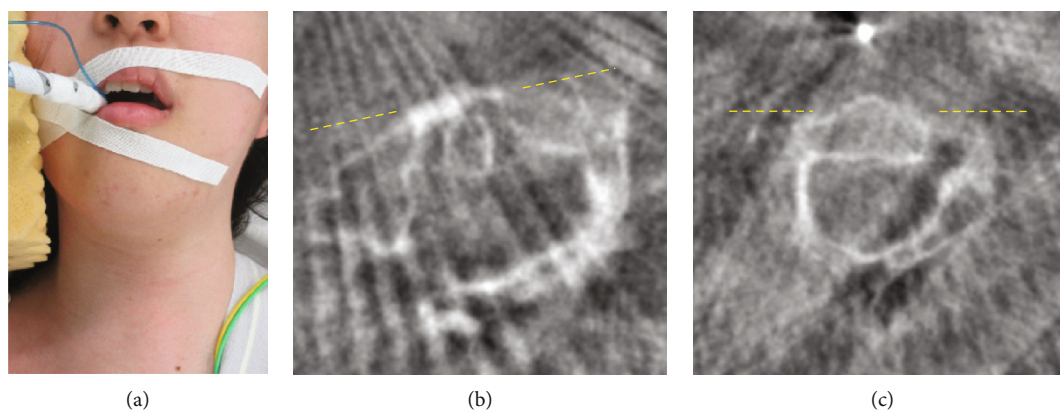


FIGURE 2: Patient's neck position (a) and CT after the first reduction (b, c). (b) Axial view of C1 and (c) axial view of C2. The C1-C2 rotatory angle decreased to 15 degrees.

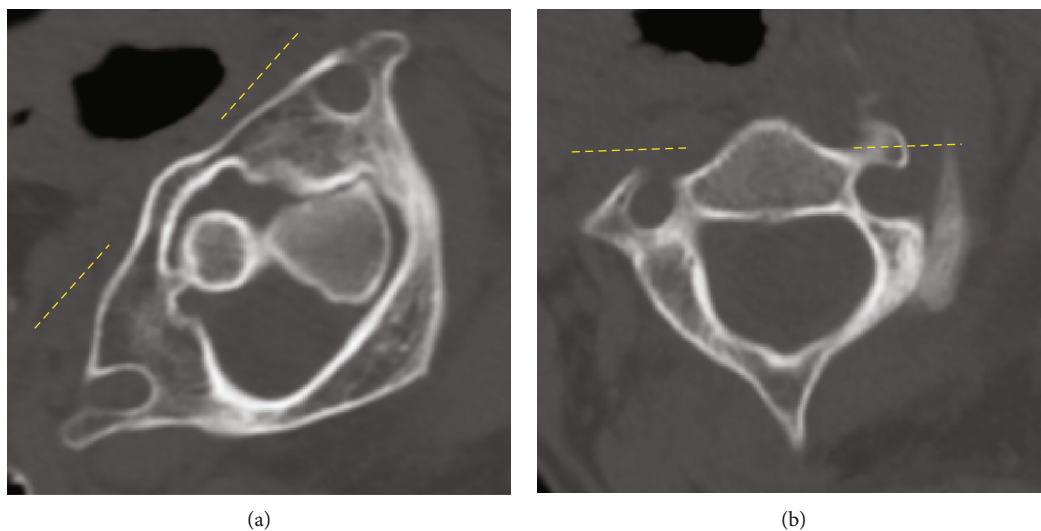


FIGURE 3: CT showing recurrence of AARF. (a) Axial view of C1 and (b) axial view of C2. The rotatory angle increased to 45 degrees.

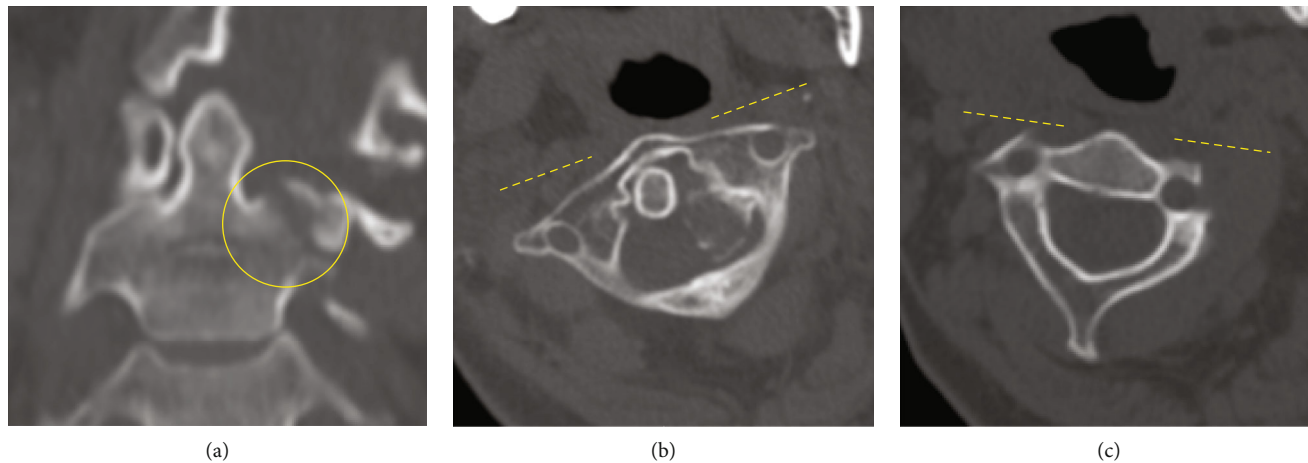


FIGURE 4: CT after the second reduction, revealing the fracture of the C2 lateral mass (yellow circle). (a) Axial view of C1, (b) axial view of C2, and (c) coronal view. Atlantoaxial rotatory subluxation remained, although the C1-C2 rotatory angle decreased to 30 degrees.

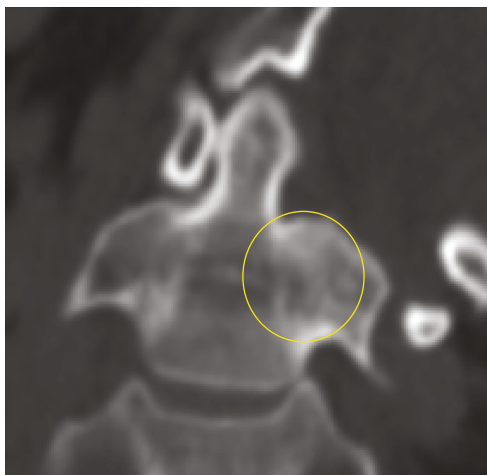


FIGURE 5: Bony union was obtained 2 months later (yellow circle).



TABLE 2: Treatment options for patients with AARF, with respect to their specific effectiveness and pros and cons [10, 24–28].

Treatment options	Effectiveness	Pros	Cons
Conservative strategies	Anti-inflammatory drugs	Limited	No mechanical invasiveness Gastrointestinal bleeding, allergic reactions
	Cervical collar	Limited	Minimum invasiveness Skin decubitus
	Halter traction Glisson traction	Limited	Low invasiveness Difficult airways access and feeding
	Skull traction	Intermediate	Intermediate invasiveness Pin site infection
Surgical strategies	Closed reduction under general anesthesia plus halo vest immobilization	High	Effective, even to chronic AARF Pin site infection
	Posterior approaches for C1-C2 fixation (various techniques: wiring, hooks, Magerl transarticular screws, and Harms technique with C1 lateral mass and C2 pars/pedicle/translaminar screws) Anterior approaches for C1-C2 fixation (various routes including lateral neck and transoral; various techniques: transarticular screws and plating)	Very high Very high	Effective in irreducible or recurrent subluxation, Fielding type 4 AARF Effective in cases with C1-C2 bony union Damage to the vertebral artery, C2 nerve root ganglion, dural tears with cerebrospinal fluid leakage Damage to the hypoglossal nerve, internal carotid artery, and soft viscera Need for nasogastric feeding

closed reduction at an early stage, before traction, halo fixation, or surgery becomes a necessary step. Optimization of medical treatment and immobilization in the hard collar may prove safer and relatively effective; nonetheless, a close clinical and radiological monitoring is recommended due to the inherently high risk of failure.

#### 4. Conclusion

AARF resulting from DICD is a rare complication of antipsychotic treatment, requiring a comprehensive assessment of pharmacologic therapy and timely radiologic workup. In this scenario, torticollis may just represent the tip of the iceberg, and only an effective multidisciplinary management can increase the chances of a satisfactory correction with closed reduction, hence avoiding the burden of more invasive treatment options.

#### Consent

The patient and her parents were informed that the data from the case would be submitted for publication and gave their consent.

#### Conflicts of Interest

The authors declare that they have no conflict of interests.

#### References

- [1] H. N. Herkowitz, S. R. Garfin, F. J. Eismont, G. R. Bell, and R. A. Balderston, *Rothman Simeone the Spine*, Elsevier Saunders, Philadelphia, PA, 6th edition, 2011.
- [2] E. M. Corner, "Rotary dislocations of the atlas," *Annals of Surgery*, vol. 45, no. 1, pp. 9–26, 1907.
- [3] G. Wortzman and F. Dewar, "Rotary fixation of the atlantoaxial joint: rotational atlantoaxial subluxation," *Radiology*, vol. 90, no. 3, pp. 479–487, 1968.
- [4] J. W. Fielding and R. Hawkins, "Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint)," *The Journal of Bone and Joint Surgery American volume*, vol. 59, no. 1, pp. 37–44, 1977.
- [5] K. M. Neal and A. S. Mohamed, "Atlantoaxial rotatory subluxation in children," *The Journal of the American Academy of Orthopaedic Surgeons*, vol. 23, no. 6, pp. 382–392, 2015.
- [6] E. C. Powell, J. R. Leonard, C. S. Olsen, D. M. Jaffe, J. Anders, and J. C. Leonard, "Atlantoaxial rotatory subluxation in children," *Pediatric Emergency Care*, vol. 33, no. 2, pp. 86–91, 2017.
- [7] O. Chechik, S. Wientroub, B. Danino, D. E. Lebel, and D. Ovidia, "Successful conservative treatment for neglected rotatory atlantoaxial dislocation," *Journal of Pediatric Orthopaedics*, vol. 33, no. 4, pp. 389–392, 2013.
- [8] D. Spiegel, S. Shrestha, P. Sitoula, N. Rendon, and J. Dormans, "Atlantoaxial rotatory displacement in children," *World Journal of Orthopedics*, vol. 8, no. 11, pp. 836–845, 2017.
- [9] D. Pang, "Atlantoaxial rotatory fixation," *Neurosurgery*, vol. 66, suppl\_3, pp. A161–A183, 2010.
- [10] K. Ishii, Y. Toyama, M. Nakamura, K. Chiba, and M. Matsumoto, "Management of chronic atlantoaxial rotatory fixation," *Spine*, vol. 37, no. 5, pp. E278–E285, 2012.
- [11] D. Pang and V. Li, "Atlantoaxial rotatory fixation: part 1—biomechanics of normal rotation at the atlantoaxial joint in children," *Neurosurgery*, vol. 55, no. 3, pp. 614–626, 2004.
- [12] S. Dalvie, A. Moore, and G. Findlay, "C1/C2 rotary subluxation due to spasmodic torticollis," *Journal of Neurology, Neurosurgery & Psychiatry*, vol. 69, no. 1, pp. 135–136, 2000.
- [13] Y. Tonomura, H. Kataoka, K. Sugie, H. Hirabayashi, H. Nakase, and S. Ueno, "Atlantoaxial rotatory subluxation associated with cervical dystonia," *Spine*, vol. 32, no. 19, pp. E561–E564, 2007.
- [14] R. M. Weigel, H.-H. Capelle, and J. K. Krauss, "Cervical dystonia in Bechterev disease resulting in atlantoaxial rotatory

- subluxation and cranio-cervical osseous fusion,” *Spine*, vol. 32, no. 25, pp. E781–E784, 2007.
- [15] S. W. Flynn, “Improvement of torticollis with quetiapine in a schizophrenia patient,” *The Canadian Journal of Psychiatry*, vol. 49, no. 2, p. 152, 2016.
- [16] S. Ryu, J. H. Yoo, J. H. Kim et al., “Tardive dyskinesia and tardive dystonia with second-generation antipsychotics in non-elderly schizophrenic patients unexposed to first-generation antipsychotics: a cross-sectional and retrospective study,” *Journal of Clinical Psychopharmacology*, vol. 35, no. 1, pp. 13–21, 2015.
- [17] D. Pang and V. Li, “Atlantoaxial rotatory fixation: part 2—new diagnostic paradigm and a new classification based on motion analysis using computed tomographic imaging,” *Neurosurgery*, vol. 57, no. 5, pp. 941–953, 2005.
- [18] P. N. van Harten, H. W. Hoek, and R. S. Kahn, “Acute dystonia induced by drug treatment,” *BMJ*, vol. 319, no. 7210, pp. 623–626, 1999.
- [19] M. F. Contarino, J. Van Den Dool, Y. Balash et al., “Clinical practice: evidence-based recommendations for the treatment of cervical dystonia with botulinum toxin,” *Frontiers in Neurology*, vol. 8, p. 35, 2017.
- [20] J. Krauss, T. Loher, T. Pohle et al., “Pallidal deep brain stimulation in patients with cervical dystonia and severe cervical dyskinesias with cervical myelopathy,” *Journal of Neurology, Neurosurgery, and Psychiatry*, vol. 72, no. 2, pp. 249–256, 2002.
- [21] E. Moro, C. LeReun, J. K. Krauss et al., “Efficacy of pallidal stimulation in isolated dystonia: a systematic review and meta-analysis,” *European Journal of Neurology*, vol. 24, no. 4, pp. 552–560, 2017.
- [22] G. Reichel, “Cervical dystonia: a new phenomenological classification for botulinum toxin therapy,” *Basal Ganglia*, vol. 1, no. 1, pp. 5–12, 2011.
- [23] D. Pang and V. Li, “Atlantoaxial rotatory fixation: part 3—a prospective study of the clinical manifestation, diagnosis, management, and outcome of children with atlantoaxial rotatory fixation,” *Neurosurgery*, vol. 57, no. 5, pp. 954–972, 2005.
- [24] S. Chibbaro, H. Cebula, S. Aldea et al., “Endonasal endoscopic odontoidectomy in ventral diseases of the craniocervical junction: results of a multicenter experience,” *World Neurosurgery*, vol. 106, pp. 382–393, 2017.
- [25] S. Chibbaro, M. Ganau, H. Cebula et al., “The endonasal endoscopic approach to pathologies of the anterior craniocervical junction: analytical review of cases treated at four European neurosurgical centres,” *Acta Neurochirurgica. Supplement*, vol. 125, pp. 187–195, 2019.
- [26] W. M. Gluf, M. H. Schmidt, and R. I. Apfelbaum, “Atlantoaxial transarticular screw fixation: a review of surgical indications, fusion rate, complications, and lessons learned in 191 adult patients,” *Journal of Neurosurgery. Spine*, vol. 2, no. 2, pp. 155–163, 2005.
- [27] P. V. Mummaneni and R. W. Haid, “Atlantoaxial fixation: overview of all techniques,” *Neurology India*, vol. 53, no. 4, pp. 408–415, 2005.
- [28] M. Payer, M. Luzi, and E. Tessitore, “Posterior atlanto-axial fixation with polyaxial C1 lateral mass screws and C2 pars screws,” *Acta Neurochirurgica*, vol. 151, no. 3, pp. 223–229, 2009.
- [29] C. C. Liao, W. W. Shen, C. C. Chang, H. Chang, and T. L. Chen, “Surgical adverse outcomes in patients with schizophrenia: a population-based study,” *Annals of Surgery*, vol. 257, no. 3, pp. 433–438, 2013.
- [30] M. E. Menendez, V. Neuhaus, A. G. Bot, D. Ring, and T. D. Cha, “Psychiatric disorders and major spine surgery: epidemiology and perioperative outcomes,” *Spine*, vol. 39, no. 2, pp. E111–E122, 2014.