

Journal of Neurological Surgery Reports

CERVICAL DIASTEMATOMYELIA: A CASE PRESENTATION AND SYSTEMATIC REVIEW

Jeff F Zhang, Oleksandr Strelko, Oleksandr Komarov, Viktoriia Kuts-Karpenko, Jonathan Forbes, Ostap Fedorko, Luke Tomycz.

Affiliations below.

DOI: 10.1055/a-2319-3444

Please cite this article as: Zhang J F, Strelko O, Komarov O et al. CERVICAL DIASTEMATOMYELIA: A CASE PRESENTATION AND SYSTEMATIC REVIEW. Journal of Neurological Surgery Reports 2024. doi: 10.1055/a-2319-3444

Conflict of Interest: The authors declare that they have no conflict of interest.

Abstract:

Diastematomyelia is a rare congenital disorder characterized by the separation of the spinal cord by an osseocartilaginous or fibrous septum. While diastematomyelia has been reported to be more common in the thoracic and lumbar regions, the true incidence of cervical diastematomyelia is currently unknown. In this study, we conducted the most comprehensive systematic review to date of all other case reports of diastematomyelia to better characterize the incidence of cervical diastematomyelia, and provide comprehensive statistics on the clinical characteristics of diastematomyelia generally. Ninety-one articles were included in our study, comprised of 252 males (27.9%) and 651 females (72.0%) (and one patient with unspecified gender). In 507 cases, the vertebral level of the diastematomyelia was described, and we recorded those levels as either cervical (n=8, 1.6%), thoracic (n=220, 43.4%), lumbar (n=277, 54.6%), or sacral (n=2, 0.4%). In 719 cases, the type of diastematomyelia was specified as either Type I (n=482, 67.0%) or Type II (n=237, 33.0%). Our study found that diastematomyelia has been reported in the cervical region in only 1.6% of cases, and we provide comprehensive data that this disorder occurs in females to males with an approximately 2.6:1 ratio, and Type I vs. Type II diastematomyelia in an estimated 2:1 ratio.

Corresponding Author:

Dr. Jeff F Zhang, Upstate University Hospital, 750 E Adams St, 13210-2399 Syracuse, United States, zhangjef@upstate.edu

Affiliations:

Jeff F Zhang, Upstate University Hospital, Syracuse, United States

Oleksandr Strelko, Loyola University Chicago Stritch School of Medicine, Neurosurgery, Maywood, United States

Oleksandr Komarov, Institute of Postgraduate Education, Bogomolets National Medical University, Neurosurgery, Kyiv, Ukraine

[...]

Luke Tomycz, Epilepsy Institute of New Jersey, Neurosurgery, Jersey City, United States



Figure 1. Localized hypertrichosis on the patient's dorsal cervical region overlying the location of the diastematomyelia.

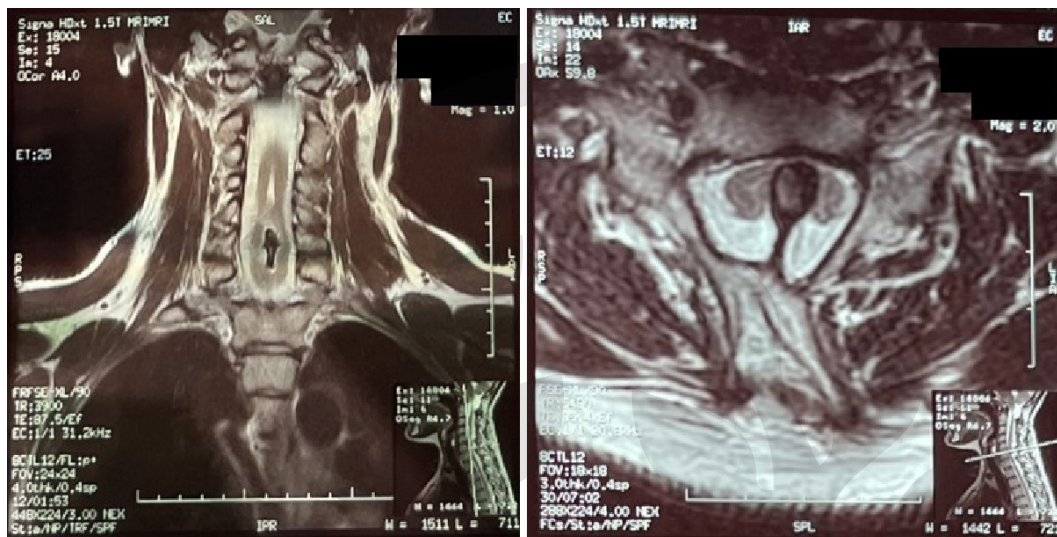


Figure 2. Coronal and axial views of the cervical vertebrae at the level of the diastematomyelia lesion.

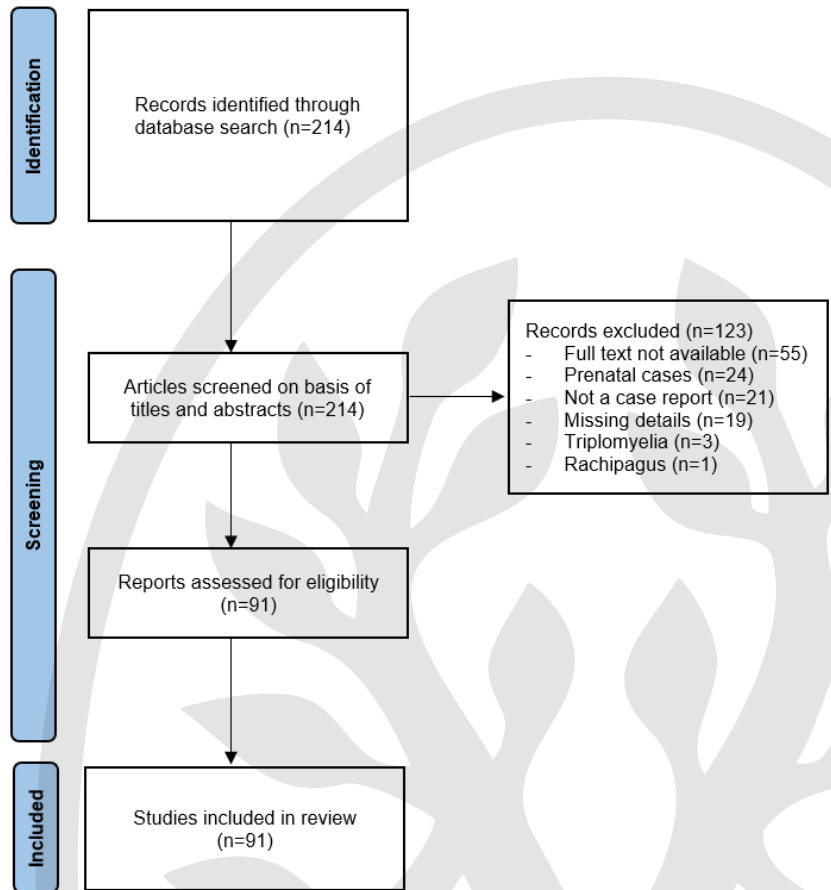


Figure 3. PRISMA flow diagram describing the search strategy used for study inclusion in this systematic review.

CERVICAL DIASTEMATOMYELIA: A CASE PRESENTATION AND SYSTEMATIC REVIEW

Jeff F Zhang MD,¹ Oleksandr Strelko BS,^{2,*} Oleksandr Komarov MD,^{3,*} Viktoriia Kuts-Karpenko MD,⁴
Jonathan A Forbes MD,⁵ Ostap Fedorko MD,⁴ Luke D Tomycz MD⁶

¹Department of Neurosurgery, SUNY Upstate Medical University, Syracuse, NY, USA

²Stritch School of Medicine, Loyola University, Maywood, IL, USA

³Institute of Postgraduate Education, Bogomolets National Medical University, Kyiv, Ukraine

⁴Clinical Municipal Communal Emergency Hospital, Lviv, Ukraine

⁵Department of Neurosurgery, University of Cincinnati College of Medicine, Cincinnati, OH, USA

⁶Epilepsy Institute of New Jersey, Jersey City, NJ, USA

*OS and OK contributed equally as co-second authors.

Correspondence

Jeff Zhang MD

Department of Neurological Surgery

750 East Adams Street

Syracuse, NY 13210

zhangjef@upstate.edu

Previous Presentation

No

Funding

The authors of this manuscript received no funding from any sources internal or external for any of the materials or findings related to this manuscript.

CRediT Authorship Statement

Conceptualization: All authors

Data curation: JFZ

Formal analysis: JFZ

Funding acquisition: N/A

Investigation: All authors

Methodology: All authors

Project administration: JAF, OF, LDT

Resources: N/A

Supervision: JAF, OF, LDT

Validation: All authors

Writing – original draft: JFZ, OS, OK

Writing – reviewing and editing: All authors

Disclosure of Financial Relationships

JFZ, OS, OK, OF, VKK, JAF, OF, LDT: None

Ethics Statement

Written and verbal consents were obtained from all patients or their healthcare proxies for all aspects related to this report and prior to any procedures which were performed.

Data Sharing

Data supporting the findings of this study will be made available by the corresponding author upon request.

Acknowledgments

We would like to thank Razom for Ukraine for providing the organizational and travel support to make this neurosurgery mission trip to Ukraine possible. We would also like to thank the neurosurgical and operative staff of the Clinical Municipal Communal Emergency Hospital and St. Nicholas Children's Hospital of Lviv for their kindness, bravery, and generosity during our time in Ukraine.

Abbreviations

CT – computed tomography
NSAID – nonsteroidal anti-inflammatory drug
SCM – spinal cord malformation
SD – standard deviation
US – United States

Abstract

Diastematomyelia is a rare congenital disorder characterized by the separation of the spinal cord by an osseocartilaginous or fibrous septum. While diastematomyelia has been reported to be more common in the thoracic and lumbar regions, the true incidence of cervical diastematomyelia is currently unknown. In this study, we conducted the most comprehensive systematic review to date of all other case reports of diastematomyelia to better characterize the incidence of cervical diastematomyelia, and provide comprehensive statistics on the clinical characteristics of diastematomyelia generally. Ninety-one articles were included in our study, comprised of 252 males (27.9%) and 651 females (72.0%) (and one patient with unspecified gender). In 507 cases, the vertebral level of the diastematomyelia was described, and we recorded those levels as either cervical (n=8, 1.6%), thoracic (n=220, 43.4%), lumbar (n=277, 54.6%), or sacral (n=2, 0.4%). In 719 cases, the type of diastematomyelia was specified as either Type I (n=482, 67.0%) or Type II (n=237, 33.0%). Our study found that diastematomyelia has been reported in the cervical region in only 1.6% of cases, and we provide comprehensive data that this disorder occurs in females to males with an approximately 2.6:1 ratio, and Type I vs. Type II diastematomyelia in an estimated 2:1 ratio.

Keywords

Diastematomyelia, neurosurgery, spinal cord malformation, spine, systematic review, Ukraine

Introduction

Diastematomyelia (also known as split cord malformation or diplomyelia) is a rare congenital disorder characterized by the separation of the spinal cord by an osseocartilaginous or fibrous septum. While the total incidence of spinal dysraphism is estimated to be 1-3 cases per 1000 live births,¹ the true incidence of diastematomyelia is unknown, though thought to occur in approximately 5% of congenital spine abnormalities.²

Split cord malformation (SCM) is classified into two types: Type I SCM in which the two hemicords are contained within two dural sacs divided by an osseous or cartilaginous septum, and Type II SCM in which a single dural tube contains both hemicords separated by a fibrous median septum.³

Diastematomyelia is usually diagnosed in childhood and associated with other congenital spine deformities in 85% of cases, such as scoliosis, tethered cord, syringomyelia, spina bifida, Chiari 2

malformation, spinal lipoma, or dermoid cyst.⁴ Type I diastematomyelia is more frequently associated with other congenital anomalies than Type II, and surgical intervention is commonly indicated for Type I patients due to symptom progression resulting from impingement of the rigid septum on the spinal cord, associated adhesions, and increasing scoliosis.⁵ Patients with Type II diastematomyelia usually only require surgery when there is significant change in scoliosis or neurological function, and symptoms in these patients tend to be milder due to the midline septum being fibrous in composition.⁵

While diastematomyelia has been reported to be more common in the thoracic and lumbar regions, the incidence of cervical diastematomyelia is thought to be extremely rare, with very few cases presented in the medical literature. In this case study, we report a patient who presented to neurosurgery clinic in Lviv Ukraine for upper extremity radicular pain during a US-Ukraine neurosurgery partnership mission (the Co-Pilot Project)⁶ and was found to have cervical diastematomyelia on imaging. We also present a thorough systematic review of all other case reports of diastematomyelia in the medical literature in order to better characterize the incidence of cervical diastematomyelia.

Case Presentation

A 31-year-old female presented to neurosurgery clinic with complaints of intermittent right shoulder pain with radiation down her arm and associated right upper extremity hypoesthesia. The patient noted that she had had these symptoms for many years, but was concerned due to increased frequency and migration of the pain from her arm to the dorsal cervical region. The patient denied any medical conditions requiring medication or any family history of connective tissue or neurological diseases. The patient noted a surgical history of spina bifida treatment at seven months of age (operative details for this surgery were unavailable), requiring two to three days of postoperative hospitalization and no complications at time of discharge.

On physical examination, the patient had noted 4/5 right hand grip and 4/5 right arm extension weakness. Lower extremity motor and neurological functions were normal, but the patient noted that her right lower extremity was approximately 3-4 cm shorter than her left lower extremity. A small tuft of hair in the patient's dorsal cervical region was appreciated on examination (**Figure 1**), from which the patient reported her muscle spasms and pain originated.

CT myelography was performed and revealed a non-contrast-enhancing bony lesion splitting the spinal cord into two asymmetric hemicords at the C6-C7 vertebral levels (**Figure 2**). No other structural anomalies were found on imaging.

Due to the patient's symptoms being well-controlled with incidental use of NSAIDs, surgical intervention was not thought to be warranted at the time of interview. The patient agreed with the course of action and was counseled to seek physical therapy and follow-up for any progression of her symptoms.

Methods

We conducted a systematic search using the Pubmed database for all full text reports in the English language describing patients with diastematomyelia. Searches were performed for all articles with "Diastematomyelia" in their title. Studies were included in our review if patients described in the case report or series had a confirmed diagnosis of diastematomyelia on imaging. All relevant studies were reviewed and information related to the number of patient(s) described, their sex and age, the spinal level of the lesion, Type I vs Type II diastematomyelia, patient treatment, and clinical outcome were recorded. The search strategy used for study selection is represented by the PRISMA flow diagram in **Figure 3**.

Results

Two-hundred and fourteen results were provided by the Pubmed database search. Of those 214 articles, 123 results were excluded due the full article text not being available (n=55), the article referring to a prenatal patient for whom a neurological examination was not possible (n=24), the article not presenting a case report or series (n=21), the article missing critical details (n=19), or the article presenting a case of triplomyelia (n=3) or diastematomyelia in a rachipagus twin (n=1). Ninety-one studies matching the inclusion criteria described in the Methods were included for data collection and further analysis. These results are shown in **Table 1**.

The 91 articles included in our study comprised reports from 904 total patients with a mean age of 23.1±22.0 years. A total of 252 males (27.9%) and 651 females (72.0%) (and one patient with unspecified gender) were included in our study. In 507 cases, the vertebral level of the diastematomyelia was described, and we recorded those levels as either cervical (n=8, 1.6%), thoracic (n=220, 43.4%), lumbar (n=277, 54.6%), or sacral (n=2, 0.4%). In 719 cases, the type of diastematomyelia was specified as either Type I (n=482, 67.0%) or Type II (n=237, 33.0%). Of 529 patients for whom follow-up data was available, 420 patients (79.4%) underwent surgical treatment for diastematomyelia, and 305 of those patients (72.6%) reported improvement in their neurological symptoms postoperatively. These findings are presented in **Table 2**.

Discussion

Consistent with previous reports of diastematomyelia found in the medical literature, the incidence of cervical diastematomyelia was found to be extremely rare, accounting for only 1.6% of all cases of diastematomyelia. The results of our comprehensive review of the literature also found that diastematomyelia has an approximately 2.5:1 predilection for females vs. males, and occurs as Type I vs Type II SCM in a 2:1 ratio.

The etiology of diastematomyelia is uncertain, but thought to be related to abnormalities in the formation of the neural tube during the fourth week of development.⁵ Adhesions between ectodermal and endodermal tissues lead to the formation of an accessory neurenteric canal in the midline of the neural tube, which results in the separation of the growing spinal cord into two hemicords as the notochord elongates rostrally.⁷ These adhesions simultaneously prevent the complete involution of fibrous septations and developmental fistulas, resulting in the formation of cysts, lipomas, and fistulas,⁸ and cause disruptions in the associated development of the surrounding vertebrae, accounting for the high proportion of comorbid spinal malformations seen in diastematomyelia patients.⁵ The accessory canal then forms the basis for the migration of mesenchymal cells which subsequently develop into the bony or cartilaginous septa seen in Type I SCM patients.⁹

While the majority of patients undergoing surgical treatment for diastematomyelia (72.6%) saw improvements in pain severity, motor function, and/or neurological symptoms, the performance of prophylactic surgery for patients with incidental findings of diastematomyelia (particularly patients with Type I SCM) on imaging is controversial.⁸ Surgical removal of an osseous septum can cause damage to the spinal cord, especially in young children, and has been reported to result in postoperative worsening of neurological symptoms in a few cases.¹⁰ Though other studies have shown that post-surgical prognoses for patients with Type I SCM are significantly improved compared to patients with Type II SCM,⁵ these qualifications of “improvement” vs. “no improvement” compared to preoperative status are frequently complicated in studies by patients who had minimal symptoms prior to surgery (especially in patients with Type II SCM). Additionally, it has been suggested that the symptoms characteristic of

diastematomyelia are related to some intrinsic myelodysplasia resulting from abnormal development rather than the presence of a bony spur in itself, as the location of a spur (resulting in the asymmetric compression of one hemicord) is not in itself predictive of the laterality or severity of symptoms.⁸ This is also corroborated by reports of patients with Type II SCM who did not have any spur detected at all on imaging but nonetheless complained of significant neurological symptoms.⁸

Limitations of our study include an inability to confirm diagnoses of diastematomyelia from an independent review of imaging in all of our included cases, and the difficulty to adequately distinguish surgical outcomes for patients with Type I vs. Type II SCM due to inconsistent reporting of results across studies and the often mixture of these two patient populations in the studies that did report surgical outcomes. The decision for surgical treatment is currently based on symptom severity or when necessary in the context of correcting concurrent spinal deformities. The establishment of clearer guidelines for surgical intervention for diastematomyelia requires further studies and trials beyond the scope of this present review.

Conclusions

Cervical diastematomyelia is an extremely rare condition, accounting for 1.6% of all cases of diastematomyelia. Clinical correlations for establishing more rigorous guidelines related to surgical intervention in cases of diastematomyelia require further studies to clarify best practices.

References

1. Rossi A. *Imaging in Spine and Spinal Cord Developmental Malformations*. Clinical Neuroradiology: The ESNR Textbook. Springer; 2018.
2. Özek MM, Cinalli G, Maixner WJ, Maixner W. *Spina Bifida: Management and Outcome*. Springer Science & Business Media; 2008.
3. Pang D, Dias MS, Ahab-Barmada M. Split cord malformation: Part I: A unified theory of embryogenesis for double spinal cord malformations. *Neurosurgery*. Sep 1992;31(3):451-80. doi:10.1227/00006123-199209000-00010
4. Ross JS, Moore KR. *Diagnostic Imaging: Spine*. Third Edition ed. Elsevier; 2016.
5. Cheng B, Li FT, Lin L. Diastematomyelia: a retrospective review of 138 patients. *J Bone Joint Surg Br*. Mar 2012;94(3):365-72. doi:10.1302/0301-620X.94B3.27897
6. Tomyzc LD, Markosian C, Kurilets I, Sr., et al. The Co-Pilot Project: An International Neurosurgical Collaboration in Ukraine. *World Neurosurg*. Mar 2021;147:e491-e515. doi:10.1016/j.wneu.2020.12.100
7. Hawryluk GWJ, Ruff CA, Fehlings MG. *Chapter 1 - Development and maturation of the spinal cord: implications of molecular and genetic defects*. vol 109. Spinal Cord Injury. Elsevier; 2012.
8. Scotti G, Musgrave MA, Harwood-Nash DC, Fitz CR, Chuang SH. Diastematomyelia in children: metrizamide and CT metrizamide myelography. *AJR Am J Roentgenol*. Dec 1980;135(6):1225-32. doi:10.2214/ajr.135.6.1225
9. Leung YL, Buxton N. Combined diastematomyelia and hemivertebra: a review of the management at a single centre. *J Bone Joint Surg Br*. Oct 2005;87(10):1380-4. doi:10.1302/0301-620X.87B10.16050
10. Goldberg C, Fenelon G, Blake NS, Dowling F, Regan BF. Diastematomyelia: a critical review of the natural history and treatment. *Spine (Phila Pa 1976)*. May-Jun 1984;9(4):367-72. doi:10.1097/00007632-198405000-00007
11. Ritchie GW, Flanagan MN. Diastematomyelia. *Can Med Assoc J*. Mar 1 1969;100(9):428-33.
12. Huang SL, He XJ, Wang KZ, Lan BS. Diastematomyelia: a 35-year experience. *Spine (Phila Pa 1976)*. Mar 15 2013;38(6):E344-9. doi:10.1097/BRS.0b013e318283f6bc

13. Kachewar SG, Sankaye SB. Diastematomyelia - a report of two cases. *J Clin Diagn Res.* Apr 2014;8(4):RE01-2. doi:10.7860/JCDR/2013/4477.4299
14. Sack AM, Khan TW. Diastematomyelia: Split Cord Malformation. *Anesthesiology.* Aug 2016;125(2):397. doi:10.1097/ALN.0000000000001021
15. Gbadamosi WA, Daftari A, Szilagyi S. Focal Diastematomyelia in an Adult: A Case Report. *Cureus.* Jun 2022;14(6):e26081. doi:10.7759/cureus.26081
16. Russell NA, Benoit BG, Joaquin AJ. Diastematomyelia in adults. A review. *Pediatr Neurosurg.* 1990;16(4-5):252-7. doi:10.1159/000120536
17. Tizard JP. Diastematomyelia. *Proc R Soc Med.* May 1957;50(5):330.
18. Saini HS, Singh M. Diastematomyelia. A case report. *Neuroradiol J.* Mar 2010;23(1):126-9. doi:10.1177/197140091002300121
19. Maebe H, Viaene A, De Muynck M. Diastematomyelia and late onset presentation: a case report of a 72-year-old woman. *Eur J Phys Rehabil Med.* Aug 2018;54(4):618-621. doi:10.23736/S1973-9087.17.04709-8
20. Hao S, Yue Z, Yu X, et al. Case report: Type I diastematomyelia with breast abnormalities and clubfoot. *Front Surg.* 2022;9:981069. doi:10.3389/fsurg.2022.981069
21. Bekki H, Morishita Y, Kawano O, Shiba K, Iwamoto Y. Diastematomyelia: a surgical case with long-term follow-up. *Asian Spine J.* Feb 2015;9(1):99-102. doi:10.4184/asj.2015.9.1.99
22. Ge CY, Hao DJ, Shan LQ. Rare Bony Diastematomyelia Associated with Intraspinal Teratoma. *World Neurosurg.* Jan 2020;133:185-187. doi:10.1016/j.wneu.2019.09.165
23. Albulescu D, Albu C, Constantin C, Stoica Z, Nicolescu I. Diastematomyelia - Imaging Findings, Case Report. *Curr Health Sci J.* Jan-Mar 2016;42(1):94-96. doi:10.12865/CHSJ.42.01.13
24. Constantinou E. A Case of Diastematomyelia. *JAMA.* Sep 21 1963;185:983-4. doi:10.1001/jama.1963.03060120093038
25. Hamidi H, Foladi N. Misdiagnosed adult presentation of diastematomyelia and tethered cord. *Radiol Case Rep.* Sep 2019;14(9):1123-1126. doi:10.1016/j.radcr.2019.06.025
26. Apostolopoulou K, Andalib A, Zaki H, deLacy P. Diastematomyelia type I associated with intramedullary lipoma and dermoid cyst. *Childs Nerv Syst.* Sep 2021;37(9):2949-2952. doi:10.1007/s00381-020-05033-3
27. Kapsalakis Z. Diastematomyelia in Two Sisters. *J Neurosurg.* Jan 1964;21:66-7. doi:10.3171/jns.1964.21.1.0066
28. Vissarionov SV, Krutelev NA, Snischuk VP, et al. Diagnosis and treatment of diastematomyelia in children: a perspective cohort study. *Spinal Cord Ser Cases.* 2018;4:109. doi:10.1038/s41394-018-0141-0
29. Hood RW, Riseborough EJ, Nehme AM, Micheli LJ, Strand RD, Neuhauser EB. Diastematomyelia and structural spinal deformities. *J Bone Joint Surg Am.* 1980;62(4):520-8.
30. Meena RK, Doddamani RS, Sharma R. Contiguous Diastematomyelia with Lipomyelomeningocele in Each Hemicord-an Exceptional Case of Spinal Dysraphism. *World Neurosurg.* Mar 2019;123:103-107. doi:10.1016/j.wneu.2018.11.225
31. Lersten M, Duhon B, Laker SR. Diastematomyelia as an Incidental Finding Lumbar on Magnetic Resonance Imaging. *PM R.* Jan 2017;9(1):95-97. doi:10.1016/j.pmrj.2016.09.005
32. Winter RB, Haven JJ, Moe JH, Lagaard SM. Diastematomyelia and congenital spine deformities. *J Bone Joint Surg Am.* Jan 1974;56(1):27-39.
33. Srinivasan ES, Mehta VA, Smith GC, Than KD, Terry AR. Klippel-Feil Syndrome with Cervical Diastematomyelia in an Adult with Extensive Cervicothoracic Fusions: Case Report and Review of the Literature. *World Neurosurg.* Jul 2020;139:274-280. doi:10.1016/j.wneu.2020.04.148
34. Kim SK, Chung YS, Wang KC, Cho BK, Choi KS, Han DH. Diastematomyelia--clinical manifestation and treatment outcome. *J Korean Med Sci.* Apr 1994;9(2):135-44. doi:10.3346/jkms.1994.9.2.135

35. Singh N, Singh DK, Kumar R. Diastematomyelia with hemimyelomeningocele: An exceptional and complex spinal dysraphism. *J Pediatr Neurosci*. Jul-Sep 2015;10(3):237-9. doi:10.4103/1817-1745.165665
36. Mamo G, Batra R, Steinig J. A Case of Diastematomyelia Presenting With Minimal Neurologic Deficits in a Middle-Aged Patient. *Cureus*. Jan 11 2021;13(1):e12621. doi:10.7759/cureus.12621
37. McNeil AG, Jose S, Rowland-Hill C. Diastematomyelia in a 3-year-old girl. *Arch Dis Child*. Jul 2018;103(7):683-684. doi:10.1136/archdischild-2017-313281
38. Hader WJ, Steinbok P, Poskitt K, Hendson G. Intramedullary spinal teratoma and diastematomyelia. Case report and review of the literature. *Pediatr Neurosurg*. Mar 1999;30(3):140-5. doi:10.1159/000028782
39. Alimli AG, Oztunali C, Boyunaga OL, et al. Diastematomyelia with the owl sign (Type I split cord malformation). *Spine J*. Oct 1 2015;15(10):e17-9. doi:10.1016/j.spinee.2015.05.025
40. Huang SL, He XJ, Xiang L, Yuan GL, Ning N, Lan BS. CT and MRI features of patients with diastematomyelia. *Spinal Cord*. Sep 2014;52(9):689-92. doi:10.1038/sc.2014.68
41. Khurram R, Ahmadi F, Poonawala R, Yasin AS. Horseshoe adrenal gland associated with type 1 diastematomyelia in an asymptomatic adult. *BJR Case Rep*. May 1 2021;7(3):20200188. doi:10.1259/bjrcr.20200188
42. Shorey WD. Diastematomyelia associated with dorsal kyphosis producing paraplegia. *J Neurosurg*. May 1955;12(3):300-5. doi:10.3171/jns.1955.12.3.0300
43. Azimi P, Mohammadi HR. Diastematomyelia Presenting With no Pain in a 53-Year-Old Man: A Case Report. *Iran Red Crescent Med J*. Jun 2013;15(6):522-5. doi:10.5812/ircmj.4195
44. Gavriiliu S, Vlad C, Georgescu I, Burnei G. Diastematomyelia in congenital scoliosis: a report of two cases. *Eur Spine J*. May 2014;23 Suppl 2:262-6. doi:10.1007/s00586-014-3218-x
45. Kansal R, Mahore A, Kukreja S. Jarcho-Levin syndrome with diastematomyelia: A case report and review of literature. *J Pediatr Neurosci*. Jul 2011;6(2):141-3. doi:10.4103/1817-1745.92843
46. English WJ, Maltby GL. Diastematomyelia in adults. *J Neurosurg*. Sep 1967;27(3):260-4. doi:10.3171/jns.1967.27.3.0260
47. Patankar T, Krishnan A, Patkar D, Armao D, Mukherji SK. Diastematomyelia and epidermoid cyst in the hemicord. *AJR Am J Roentgenol*. Jun 2000;174(6):1793-4. doi:10.2214/ajr.174.6.1741793
48. Gan YC, Sgouros S, Walsh AR, Hockley AD. Diastematomyelia in children: treatment outcome and natural history of associated syringomyelia. *Childs Nerv Syst*. May 2007;23(5):515-9. doi:10.1007/s00381-006-0205-5
49. Yamanaka T, Hashimoto N, Sasajima H, Mineura K. A case of diastematomyelia associated with myeloschisis in a hemicord. *Pediatr Neurosurg*. Nov 2001;35(5):253-6. doi:10.1159/000050431
50. Beyerl BD, Ojemann RG, Davis KR, Hedley-Whyte ET, Mayberg MR. Cervical diastematomyelia presenting in adulthood. Case report. *J Neurosurg*. Mar 1985;62(3):449-53. doi:10.3171/jns.1985.62.3.0449
51. Sandhu J, Gupta SK, Katha M. An unusual Case of Faun Tail Nevus with Aplasia Cutis, Dermo-fascial Sinus Defect, Diastematomyelia, and Spinal cord Syrinx. *Indian J Dermatol*. May-Jun 2021;66(3):322-324. doi:10.4103/ijd.IJD_816_19
52. Chembolli L. Faun Tail Overlying Spinal Dysraphism (Diastematomyelia) at the Mid Thoracic Level: Cosmetic Improvement Achieved with Diode Laser Epilation. *Indian J Dermatol*. Nov-Dec 2015;60(6):638. doi:10.4103/0019-5154.169165
53. Elmaci I, Dacinar A, Ozgen S, Ekinci G, Pamir MN. Diastematomyelia and spinal teratoma in an adult. Case report. *Neurosurg Focus*. Jan 15 2001;10(1):ecp2. doi:10.3171/foc.2001.10.1.10
54. Xu L, Ma C, Shen S, Duan H, Li X. A heterozygous mutation in the ALPL gene in an adolescent with Chiari malformation type I accompanied by scoliosis, tethered cord and diastematomyelia. *Acta Neurol Belg*. Jan 28 2023;doi:10.1007/s13760-023-02197-y
55. Zaleska-Dorobisz U, Bladowska J, Biel A, Palka LW, Holownia D. MRI diagnosis of diastematomyelia in a 78-year-old woman: Case report and literature review. *Pol J Radiol*. Apr 2010;75(2):82-7.

56. Sheehan JP, Sheehan JM, Lopes MB, Jane JA, Sr. Thoracic diastematomyelia with concurrent intradural epidermoid spinal cord tumor and cervical syrinx in an adult. Case report. *J Neurosurg.* Sep 2002;97(2 Suppl):231-4. doi:10.3171/spi.2002.97.2.0231
57. Kanbur NO, Guner P, Derman O, Akalan N, Cila A, Kutluk T. Diastematomyelia: a case with familial aggregation of neural tube defects. *ScientificWorldJournal.* Sep 21 2004;4:847-52. doi:10.1100/tsw.2004.140
58. Tubbs RS, Smyth MD, Dure LS, Oakes WJ. Exclusive lower extremity mirror movements and diastematomyelia. *Pediatr Neurosurg.* May-Jun 2004;40(3):132-5. doi:10.1159/000079856
59. Shivapathasundram G, Stoodley MA. Use of a synthetic dural substitute to prevent ventral retethering in the management of diastematomyelia. *J Clin Neurosci.* Apr 2012;19(4):578-81. doi:10.1016/j.jocn.2011.08.018
60. Parmar H, Patkar D, Shah J, Maheshwari M. Diastematomyelia with terminal lipomyelocystocele arising from one hemicord: case report. *Clin Imaging.* Jan-Feb 2003;27(1):41-3. doi:10.1016/s0899-7071(02)00522-3
61. Senkoylu A, Cetinkaya M, Aktas E, Cetin E. Excision and short segment fusion of a double ipsilateral lumbar hemivertebrae associated with a diastematomyelia and fixed pelvic obliquity. *Acta Orthop Traumatol Turc.* Mar 2019;53(2):160-164. doi:10.1016/j.aott.2019.01.001
62. Tsitsopoulos P, Rizos C, Isaakidis D, Liapi G, Zymaris S. Coexistence of spinal intramedullary teratoma and diastematomyelia in an adult. *Spinal Cord.* Oct 2006;44(10):632-5. doi:10.1038/sj.sc.3101886
63. Pettorini BL, Massimi L, Cianfoni A, Paternoster G, Tamburini G, Di Rocco C. Thoracic lipomeningocele associated with diastematomyelia, tethered spinal cord, and hydrocephalus. Case report. *J Neurosurg.* May 2007;106(5 Suppl):394-7. doi:10.3171/ped.2007.106.5.394
64. Lewandowski KU, Rachlin JR, Glazer PA. Diastematomyelia presenting as progressive weakness in an adult after spinal fusion for adolescent idiopathic scoliosis. *Spine J.* Jan-Feb 2004;4(1):116-9. doi:10.1016/j.spinee.2003.08.028
65. Ross GW, Swanson SA, Perentes E, Urich H. Ectopic midline spinal ganglion in diastematomyelia: a study of its connections. *J Neurol Neurosurg Psychiatry.* Sep 1988;51(9):1231-4. doi:10.1136/jnnp.51.9.1231
66. Porensky P, Muro K, Ganju A. Adult presentation of spinal dysraphism and tandem diastematomyelia. *Spine J.* Sep-Oct 2007;7(5):622-6. doi:10.1016/j.spinee.2006.08.006
67. Filippi CG, Andrews T, Gonyea JV, Linnell G, Cauley KA. Magnetic resonance diffusion tensor imaging and tractography of the lower spinal cord: application to diastematomyelia and tethered cord. *Eur Radiol.* Sep 2010;20(9):2194-9. doi:10.1007/s00330-010-1797-4
68. Senel E, Tiryaki T, Atayurt H, Cansu A, Guc T. Lumbo-costovertebral syndrome with diastematomyelia. *Pediatr Int.* Aug 2008;50(4):600-2. doi:10.1111/j.1442-200X.2008.02676.x
69. Shen J, Zhang J, Feng F, Wang Y, Qiu G, Li Z. Corrective Surgery for Congenital Scoliosis Associated with Split Cord Malformation: It May Be Safe to Leave Diastematomyelia Untreated in Patients with Intact or Stable Neurological Status. *J Bone Joint Surg Am.* Jun 1 2016;98(11):926-36. doi:10.2106/JBJS.15.00882
70. Sharma A, Sharma R, Goyal M, Vashisht S, Berry M. Diastematomyelia associated with intramedullary tumour in a hemicord: a report of two cases. *Australas Radiol.* May 1997;41(2):185-7.
71. Sgouros S. Acquired Chiari I malformation in a child with corrected diastematomyelia disappeared after thickened filum division. *Pediatr Neurosurg.* 2010;46(5):402-5. doi:10.1159/000323423
72. Wenger M, Hauswirth CB, Brodhage RP. Undiagnosed adult diastematomyelia associated with neurological symptoms following spinal anaesthesia. *Anaesthesia.* Aug 2001;56(8):764-7. doi:10.1046/j.1365-2044.2001.01916.x
73. Kilickesmez O, Barut Y, Tasmiroglu E. Expanding occult intrasacral meningocele associated with diastematomyelia and multiple vertebral anomalies. Case report. *J Neurosurg.* Aug 2004;101(1 Suppl):108-11. doi:10.3171/ped.2004.101.2.0108

74. Kaminker R, Fabry J, Midha R, Finkelstein JA. Split cord malformation with diastematomyelia presenting as neurogenic claudication in an adult: a case report. *Spine (Phila Pa 1976)*. Sep 1 2000;25(17):2269-71. doi:10.1097/00007632-200009010-00021
75. Ak H, Atalay T, Gulsen I. The association of the epidermoid cyst of the filum terminale, intradural spinal lipoma, tethered cord, dermal sinus tract, and type I diastematomyelia in a child. *World Neurosurg*. Dec 2014;82(6):e836-7. doi:10.1016/j.wneu.2014.08.040
76. Ohwada T, Okada K, Hayashi H. Thoracic myelopathy caused by cervicothoracic diastematomyelia. A case report. *J Bone Joint Surg Am*. Feb 1989;71(2):296-9.
77. Morelli C, Schalick WO, 3rd. Diastematomyelia presenting in adulthood as back pain. *Am J Phys Med Rehabil*. Sep 2013;92(9):838. doi:10.1097/PHM.0b013e3182241837
78. Macht S, Chapot R, Bieniek F, Hanggi D, Turowski B. Unique sacral location of an arteriovenous fistula of the filum terminale associated with diastematomyelia and lowered spinal cords. *Neuroradiology*. May 2012;54(5):517-9. doi:10.1007/s00234-011-0899-2
79. Kanagaraju V, Chhabra HS, Srivastava A, et al. A case of severe and rigid congenital thoracolumbar lordoscoliosis with diastematomyelia presenting with type 2 respiratory failure: managed by staged correction with controlled axial traction. *Eur Spine J*. Oct 2016;25(10):3034-3041. doi:10.1007/s00586-014-3624-0
80. Boussaadani Soubai R, Tahiri L, Sqalli Houssaini G, Mansouri S, Harzy T. Adult presentation of diastematomyelia: a case report. *Joint Bone Spine*. Oct 2011;78(5):529-30. doi:10.1016/j.jbspin.2011.03.025
81. Armstrong DJ, McCormick D, O'Longain D. Previously undiagnosed diastematomyelia with bony spur as a cause of back pain in a 49-year-old patient with known psoriatic arthritis. *Rheumatology (Oxford)*. Feb 2017;56(2):238. doi:10.1093/rheumatology/kew359
82. Giordano N, Cicone C, Hadilaksono MG, Agarwal S, Kifle G. Poster 375 Adult Onset Lumbar Radiculopathy Secondary to Type II Diastematomyelia: A Case Report. *PM R*. Sep 2016;8(9S):S283. doi:10.1016/j.pmrj.2016.07.302
83. Sharma MC, Sarat Chandra P, Goel S, Gupta V, Sarkar C. Primary lumbosacral Wilms tumor associated with diastematomyelia and occult spinal dysraphism. A report of a rare case and a short review of literature. *Childs Nerv Syst*. Mar 2005;21(3):240-3. doi:10.1007/s00381-004-0989-0
84. Sedzimir CB, Roberts JR, Occleshaw JV. Massive diastematomyelia without cutaneous dysraphism. *Arch Dis Child*. May 1973;48(5):400-2. doi:10.1136/adc.48.5.400
85. Callari G, Arrigo A. Diastematomyelia in adults. A report of two cases incidentally discovered. *Neuroradiol J*. Aug 29 2009;22(4):448-51. doi:10.1177/197140090902200415
86. Kramer JL, Dvorak M, Curt A. Thoracic disc herniation in a patient with tethered cord and lumbar syringomyelia and diastematomyelia: magnetic resonance imaging and neurophysiological findings. *Spine (Phila Pa 1976)*. Jun 15 2009;34(14):E484-7. doi:10.1097/BRS.0b013e31819211c9
87. Roche J, Vignaendra D. Midline septa in the lumbo-sacral thecal sac: acquired abnormality or developmental anomaly? The equivalent of diastematomyelia occurring below the spinal cord? *Australas Radiol*. Dec 2006;50(6):553-62. doi:10.1111/j.1440-1673.2006.01652.x
88. Uzumcugil A, Cil A, Yazici M, et al. The efficacy of convex hemiepiphysiodesis in patients with iatrogenic posterior element deficiency resulting from diastematomyelia excision. *Spine (Phila Pa 1976)*. Apr 15 2003;28(8):799-805.
89. Burnei G, Gavriiliu TS, Vlad C, Japie EM, Ghita RA. L3-L5 teratological spondylolysis with diastematomyelia and L4 radicular syndrome followed by spondyloschisis without myelomeningocele due to somatoarcuate shifting. *Spine J*. Jan 1 2015;15(1):202-4. doi:10.1016/j.spinee.2014.09.006
90. Hung PC, Wang HS, Lui TN, Wong AM. Sonographic findings in a neonate with diastematomyelia and a tethered spinal cord. *J Ultrasound Med*. Sep 2010;29(9):1357-60. doi:10.7863/jum.2010.29.9.1357
91. Turgut M, Doger FK. A case of diastematomyelia associated with hamartoma masquerading as meningocele in the newborn infant. *Pediatr Neurosurg*. 2008;44(1):85-7. doi:10.1159/000110671

92. Bale PM. A congenital intraspinal gastroenterogenous cyst in diastematomyelia. *J Neurol Neurosurg Psychiatry*. Dec 1973;36(6):1011-7. doi:10.1136/jnnp.36.6.1011
93. Korinth MC, Kapser A, Nolte K, Gilsbach JM. Cervical diastematomyelia associated with an intradural epidermoid cyst between the hemicords and multiple vertebral body anomalies. *Pediatr Neurosurg*. Sep-Oct 2004;40(5):253-6. doi:10.1159/000082303
94. Mendez JC, Prieto MA, Lanciego C. Percutaneous vertebroplasty in a patient with type I split cord malformation (diastematomyelia). *Cardiovasc Intervent Radiol*. May 2009;32(3):608-10. doi:10.1007/s00270-009-9560-4
95. Yamada S, Mandybur GT, Thompson JR. Dorsal midline proboscis associated with diastematomyelia and tethered cord syndrome. Case report. *J Neurosurg*. Oct 1996;85(4):709-12. doi:10.3171/jns.1996.85.4.0709
96. Lourie H, Bierny JP. Diastematomyelia with two spurs and intradural neural crest elements. Case report. *J Neurosurg*. Feb 1970;32(2):248-51. doi:10.3171/jns.1970.32.2.0248
97. Ugarte N, Gonzalez-Crussi F, Sotelo-Avila C. Diastematomyelia associated with teratomas. Report of two cases. *J Neurosurg*. Nov 1980;53(5):720-5. doi:10.3171/jns.1980.53.5.0720
98. Okada K, Fuji T, Yonenobu K, Ono K. Cervical diastematomyelia with a stable neurological deficit. Report of a case. *J Bone Joint Surg Am*. Jul 1986;68(6):934-7.
99. Azhar MM, Winter RB, Dunn MB. Congenital spine deformity, congenital stenosis, diastematomyelia, and tight filum terminale in a workmen's compensation patient: a case report. *Spine (Phila Pa 1976)*. Mar 15 1996;21(6):770-4. doi:10.1097/00007632-199603150-00024

Table 1: Systematic Review of Cases of Diastematomyelia



Author	n	Sex/Age	Spinal Level	Type	Treatment	Outcome
Ritchie and Flanagan 1969 ¹¹	8	M: 2, F: 6 1 week-9 years	T: 4 L: 3 U: 1	II: 8	Surgery: 8	Improvement: 2/8
Huang et al. 2013 ¹²	156	M: 47, F: 109 Mean=4.5 years	C: 2 T: 82 L: 72	I: 123 II: 33	Surgery: 121 Non-Surgical: 35	Improvement: (I: 96/123), (II: 0/33)
Kachewar and Sankaye 2014 ¹³	2	M: 2 (17 years and 1 year)	T: 1 L: 1	U	U	U
Sack and Khan 2016 ¹⁴	1	F: 1 (29 years)	T: 1	II: 1	U	U
Gbadamosi, Daftari, Szilagyi 2022 ¹⁵	1	F: 1 (U)	L: 1	I: 1	Non-Surgical	U
Russell, Benoit, Joaquin 1990 ¹⁶	45	M: 12, F: 33 Mean=37.8 years	U	U	Surgical: 24 Non-Surgical: 21	Improvement: (Surgical: 23/24)
Tizard 1957 ¹⁷	1	F: 1 (3 years)	C: 1	U	Non-Surgical	U
Saini and Singh 2009 ¹⁸	1	M: 1 (22 days)	L: 1	I: 1	U	U
Maebe et al. 2018 ¹⁹	1	F: 1 (72 years)	L: 1	I: 1	Non-Surgical	U
Hao et al. 2022 ²⁰	1	F: 1 (18 years)	L: 1	I: 1	Surgical	Improvement
Bekki et al. 2015 ²¹	1	F: 1 (14 years)	T: 1	I: 1	Surgical	Improvement
Ge, Hao, Shan 2020 ²²	1	M: 1 (36 years)	L: 1	I: 1	Surgical	Improvement
Albulescu et al. 2016 ²³	1	U: 1 (45 years)	L: 1	I: 1	U	U
Cheng, Li, Lin 2012 ⁵	138	M: 34, F: 104 Mean=15.7 years	U	I: 106 II: 32	Surgical: 112 Non-Surgical: 26	Improvement: (Surgical: I: 91/96), (Surgical: II: 8/16)
Constantinou 1963 ²⁴	1	F: 1 (23 years)	L: 1	I: 1	Non-Surgical	U
Hamidi and Foladi 2019 ²⁵	1	M: 1 (48 years)	L: 1	I: 1	Non-Surgical	U
Apostolopoulou 2021 ²⁶	1	F: 1 (5 years)	L: 1	I: 1	Surgical	Improvement
Kapsalakis 1964 ²⁷	2	F: 2 (6 years and 5 years)	L: 2	I: 2	Surgical	Improvement: 1/2
Vissarionov et al. 2018 ²⁸	20	M: 8, F: 12 Mean=9.2 years	T:15 L: 5	U	Surgical: 17 Non-Surgical: 3	Improvement: (Surgical: 17/17)
Hood et al. 1980 ²⁹	60	M: 13, F: 47 Mean=4.7 years	T: 24 L: 36	U	Surgical: 51 Non-Surgical: 9	Improvement: (Surgical: 20/51)
Meena, Doddamani,	1	F: 1 (15	L: 1	I: 1	Surgical	No improvement

Sharma 2018 ³⁰		months)				
Lersten, Duhon, Laker 2017 ³¹	1	F: 1 (50 years)	L: 1	I: 1	Non-Surgical	U
Winter et al. 1974 ³²	27	M: 6, F: 21 Mean=6.5 years	T: 8 L: 19	U	Surgical: 22 Non-Surgical: 5	Improvement: (Surgical: 5/19)
Srinivasan et al. 2020 ³³	1	F: 1 (55 years)	C: 1	II: 1	Non-Surgical	Improvement
Kim et al. 1994 ³⁴	5	M: 3, F: 2 Mean=14.8 years	L: 5	I: 5	Surgical: 5	Improvement: 4/5
Singh, Singh, Kumar 2015 ³⁵	1	F: 1 (3 months)	T: 1	I: 1	Surgical	U
Mamo, Batra, Steinig 2021 ³⁶	1	M: 1 (50 years)	T: 1	I: 1	Non-Surgical	U
McNeil, Jose, Rowland-Hill 2018 ³⁷	1	F: 1 (3 years)	L: 1	I: 1	U	U
Hader et al. 1999 ³⁸	1	F: 1 (16 years)	L: 1	I: 1	Surgical	U
Alimli et al. 2015 ³⁹	1	F: 1 (4 years)	T: 1	I: 1	Surgical	U
Huang et al. 2014 ⁴⁰	82	M: 17, F: 65 Median=6 years	T: 50 L: 32	I: 82	U	U
Khurram et al. 2021 ⁴¹	1	M: 1 (38 years)	L: 1	I: 1	U	U
Shorey 1955 ⁴²	1	M: 1 (12 years)	L: 1	I: 1	Surgical	Improvement
Azimi and Mohammadi 2013 ⁴³	1	M: 1 (53 years)	L: 1	I: 1	Surgical	Improvement
Gavriliu et al. 2014 ⁴⁴	2	M: 1 (12 years), F: 1 (7 years)	T: 1 L: 1	I: 2	Surgical	Improvement: 1/2
Scotti et al. 1980 ⁸	21	M: 8, F: 13 Mean=7.5 years	L: 21	I: 5 II: 15	Surgical: 15 Non-Surgical: 6	U
Kansal, Mahore, Kukreja 2011 ⁴⁵	1	M: 1 (1.5 years)	L: 1	I: 1	Surgical	No improvement
English and Malthy 1967 ⁴⁶	2	M: 1 (48 years), F: 1 (32 years)	L: 2	U	Surgical: 1 Non-Surgical: 1	Improvement: (Surgical: 0/1) No improvement: (Non-Surgical: U)
Patankar et al. 2000 ⁴⁷	1	F: 1 (5 years)	T: 1	I: 1	Surgical	Improvement
Gan et al. 2007 ⁴⁸	17	M: 8, F: 9 Mean=3.4 years	T: 5 L: 12	I: 17	Surgical: 17	Improvement: 5/17
Yamanaka et al. 2001 ⁴⁹	1	U: 1 (6 days)	L: 1	I: 1	Surgical	Improvement
Beyerl et al. 1985 ⁵⁰	1	M: 1 (34 years)	C: 1	I: 1	Surgical	Improvement
Sandhu, Gupta, Katha 2021 ⁵¹	1	M: 1 (25 years)	T: 1	II: 1	Non-Surgical	Improvement
Chembolli 2015 ⁵²	1	F: 1 (16 years)	T: 1	I: 1	Surgical	Improvement
Elmaci et al. 2001 ⁵³	1	M: 1 (42	L:1	I: 1	Surgical	Improvement

		years)				
--	--	--------	--	--	--	--



Xu et al. 2023 ⁵⁴	1	F: 1 (17 years)	L: 1	I: 1	Surgical	Improvement
Zaleska-Dorobisz et al. 2010 ⁵⁵	1	F: 1 (78 years)	L: 1	I: 1	U	U
Sheehan, Sheehan, Lopes 2002 ⁵⁶	1	F: 1 (38 years)	T: 1	I: 1	Surgical	Improvement
Kanbur et al. 2004 ⁵⁷	1	M: 1 (12 years)	L: 1	I: 1	Non-Surgical	No improvement
Tubbs et al. 2004 ⁵⁸	1	F: 1 (18 years)	T: 1	I: 1	Surgical	No improvement
Shivapathasundram and Stoodley 2012 ⁵⁹	1	F: 1 (8 years)	T: 1	I: 1	Surgical	Improvement
Parmar et al. 2003 ⁶⁰	1	F: 1 (34 years)	L: 1	I: 1	Surgical	Improvement
Senkoylu et al. 2019 ⁶¹	1	F: 1 (4 years)	L: 1	I: 1	Surgical	Improvement
Tsitsopoulos et al. 2006 ⁶²	1	F: 1 (44 years)	L: 1	I: 1	Non-Surgical	No improvement
Pettorini et al. 2007 ⁶³	1	M: 1 (2 years)	T: 1	I: 1	Surgical	Improvement
Lewandowski, Rachlin, Glazer 2004 ⁶⁴	1	F: 1 (44 years)	L: 1	I: 1	Surgical	Improvement
Ross et al. 1988 ⁶⁵	1	M: 1 (63 years)	S: 1	I: 1	Non-Surgical	No improvement
Porensky, Muro, Ganju 2007 ⁶⁶	1	F: 1 (54 years)	T: 1 L: 1	I: 2	Surgical	Improvement
Filippi et al. 2010 ⁶⁷	3	M: 1 (67 years), F: 2 (53 and 49 years)	T: 1 L: 2	I: 1 II: 2	U	U
Senel et al. 2008 ⁶⁸	1	F: 1 (14 months)	T: 1	I: 1	Surgical	U
Shen et al. 2016 ⁶⁹	21 4	M: 61, F: 153 Mean=14.2 years	U	I: 73 II: 141	Surgical: 214	U
Sharma et al. 1997 ⁷⁰	1	M: 1 (15 years), F: 1 (9 years)	T: 1 L: 1	I: 2	Surgical: 2	U
Sgouros 2010 ⁷¹	1	F: 1 (3 years)	L: 1	I: 1	Surgical	Improvement
Wenger, Hauswirth, Brodhage 2001 ⁷²	1	F: 1 (38 years)	L: 1	II: 1	Non-Surgical	Improvement
Kilickesmez, Barut, Tasmiroglu 2004 ⁷³	1	F: 1 (7 years)	L: 1	I: 1	Surgical	Improvement
Kaminker et al. 2000 ⁷⁴	1	M: 1 (38 years)	L: 1	I: 1	Surgical	Improvement
Ak, Atalay, Gulsen 2014 ⁷⁵	1	F: 1 (10 years)	L: 1	I: 1	Surgical	Improvement
Ohwada, Okada, Hayashi 1989 ⁷⁶	1	M: 1 (29 years)	C: 1	I: 1	Surgical	Improvement
Morelli and Shalick 2011 ⁷⁷	1	F: 1 (29 years)	L: 1	I: 1	Surgical	No improvement
Macht et al. 2012 ⁷⁸	1	M: 1 (57 years)	L: 1	I: 1	Non-Surgical	U
Kanagaraju et al. 2016 ⁷⁹	1	F: 1 (15 years)	L: 1	I: 1	Non-Surgical	Improvement
Boussaandani et al. 2011 ⁸⁰	1	F: 1 (33 years)	L: 1	I: 1	Non-Surgical	U
Armstrong, McCormick,	1	F: 1 (49 years)	L: 1	I: 1	Non-Surgical	U

O'Longain 2016 ⁸¹						
Giordano et al. 2016 ⁸²	1	F: 1 (43 years)	T: 1	II: 1	U	U
Sharma et al. 2005 ⁸³	1	F: 1 (18 months)	L: 1	I: 1	Surgical	Improvement
Sedzimir, Roberts, Occleshaw 1973 ⁸⁴	1	M: 1 (22 months)	L: 1	I: 1	Surgical	Improvement
Callari and Arrigo 2009 ⁸⁵	2	F: 2 (80 and 59 years)	L: 2	U	Non-Surgical	Improvement: 2/2
Kramer, Dvorak, Curt 2009 ⁸⁶	1	F: 1 (54 years)	L: 1	I: 1	Surgical	No improvement
Roche and Vignaendra 2006 ⁸⁷	8	M: 3, F: 5 Mean=53.8 years	L: 7 S: 1	U	U	U
Uzumcugil et al. 2003 ⁸⁸	18	M: 2, F: 16 Mean=20 months	T: 9 L: 9	U	Surgical	U
Burnei et al. 2015 ⁸⁹	1	F: 1 (18 years)	L: 1	I: 1	Surgical	Improvement
Hung et al. 2010 ⁹⁰	1	F: 1 (2 days)	L: 1	I: 1	Surgical	U
Turgut and Doger 2008 ⁹¹	1	M: 1 (1 day)	L: 1	I: 1	Surgical	U
Bale 1973 ⁹²	1	F: 1 (6 days)	T: 1	I: 1	Surgical	U
Korinth et al. 2004 ⁹³	1	M: 1 (2 years)	C: 1	I: 1	Surgical	No improvement
Mendez, Prieto, Lanciego 2009 ⁹⁴	1	F: 1 (88 years)	L: 1	I: 1	Non-Surgical	Improvement
Yamada, Mandybur, Thompson 1996 ⁹⁵	1	F: 1 (2 years)	L: 1	I: 1	Surgical	Improvement
Lourie and Bierny 1970 ⁹⁶	1	F: 1 (7 years)	T: 1	I: 1	Surgical	U
Ugarte, Gonzalez-Crussi, Sotelo-Avila 1980 ⁹⁷	2	F: 2 (1 day and 1 day)	T: 2	U	Surgical	Died: 2/2
Okada et al. 1986 ⁹⁸	1	M: 1 (19 years)	C: 1	I: 1	Non-Surgical	U
Azhar, Winter, Dunn 1996 ⁹⁹	1	M: 1 (35 years)	L: 1	I: 1	Surgical	Improvement

Table 1. Systematic review of case reports and series describing patients with diastematomyelia. “n” number of patients described in the study, the sex (M = male, F = female, U = unknown) and age of included patients, the spinal location of the lesion (C = cervical, T = thoracic, L = lumbar, U = unknown), Type I vs. Type II spinal cord malformation, treatment, and clinical outcome were recorded.

Table 2: Patient Demographics

	Population (n=904)
Age (mean ± SD)	23.1 ± 22.0
Gender	
Male (n, %)	252 (27.9)
Female (n, %)	651 (72.0)
Unspecified (n, %)	1 (0.1)
Vertebral Level	507 (56.1)
Cervical (n, %)	8 (1.6)
Thoracic (n, %)	220 (43.4)
Lumbar (n, %)	277 (54.6)
Sacral (n, %)	2 (0.4)
Pang Criteria	719 (79.5)
Type I (n, %)	482 (67.0)
Type II (n, %)	237 (33.0)
Surgical Treatment	420 (79.4)
Improvement (n, %)	305 (72.6)
No Improvement (n, %)	115 (27.4)

Table 2: Patient Demographics. Determinations of clinical improvement following treatment were calculated from the n=529 patients for whom follow-up data were provided. SD, standard deviation