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## CERVICAL DIASTEMATOMYELIA: A CASE PRESENTATION AND SYSTEMATIC REVIEW

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### 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.

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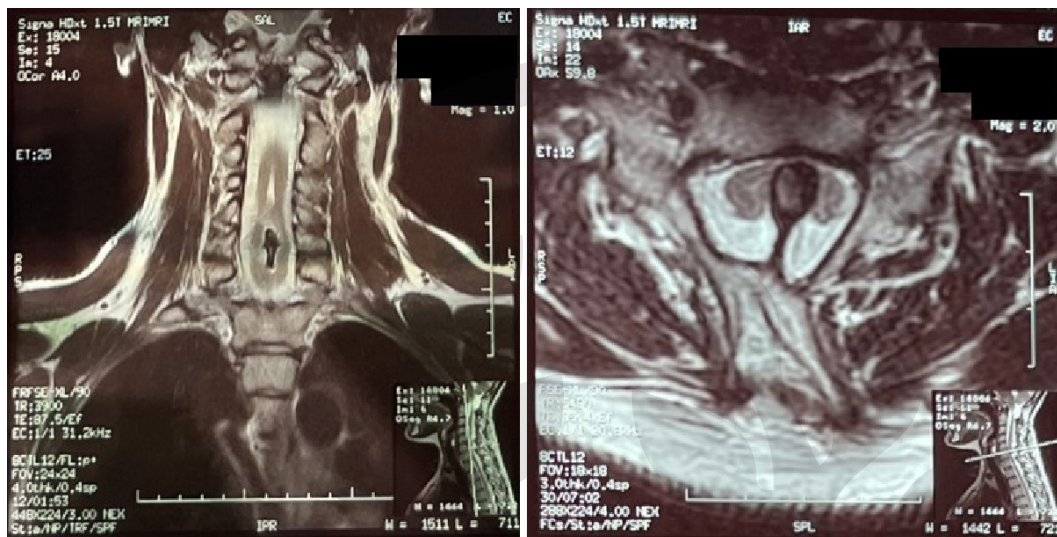
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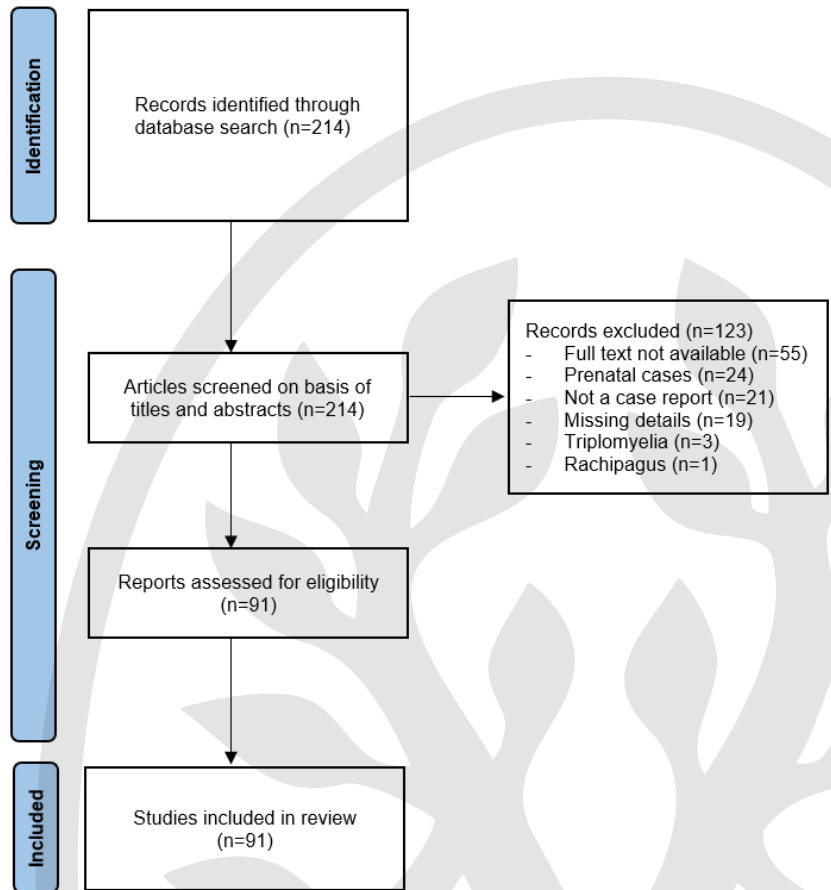
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**Figure 1.** Localized hypertrichosis on the patient's dorsal cervical region overlying the location of the diastematomyelia.



**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

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### 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.

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### **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,<sup>1</sup> the true incidence of diastematomyelia is unknown, though thought to occur in approximately 5% of congenital spine abnormalities.<sup>2</sup>

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.<sup>3</sup>

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.<sup>4</sup> 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.<sup>5</sup> 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.<sup>5</sup>

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)<sup>6</sup> 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.<sup>5</sup> 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.<sup>7</sup> These adhesions simultaneously prevent the complete involution of fibrous septations and developmental fistulas, resulting in the formation of cysts, lipomas, and fistulas,<sup>8</sup> and cause disruptions in the associated development of the surrounding vertebrae, accounting for the high proportion of comorbid spinal malformations seen in diastematomyelia patients.<sup>5</sup> 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.<sup>9</sup>

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.<sup>8</sup> 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.<sup>10</sup> 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,<sup>5</sup> 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.<sup>8</sup> 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.<sup>8</sup>

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.

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**Table 1: Systematic Review of Cases of Diastematomyelia**



Author	n	Sex/Age	Spinal Level	Type	Treatment	Outcome
Ritchie and Flanagan 1969 <sup>11</sup>	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 <sup>12</sup>	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 <sup>13</sup>	2	M: 2 (17 years and 1 year)	T: 1 L: 1	U	U	U
Sack and Khan 2016 <sup>14</sup>	1	F: 1 (29 years)	T: 1	II: 1	U	U
Gbadamosi, Daftari, Szilagyi 2022 <sup>15</sup>	1	F: 1 (U)	L: 1	I: 1	Non-Surgical	U
Russell, Benoit, Joaquin 1990 <sup>16</sup>	45	M: 12, F: 33 Mean=37.8 years	U	U	Surgical: 24 Non-Surgical: 21	Improvement: (Surgical: 23/24)
Tizard 1957 <sup>17</sup>	1	F: 1 (3 years)	C: 1	U	Non-Surgical	U
Saini and Singh 2009 <sup>18</sup>	1	M: 1 (22 days)	L: 1	I: 1	U	U
Maebe et al. 2018 <sup>19</sup>	1	F: 1 (72 years)	L: 1	I: 1	Non-Surgical	U
Hao et al. 2022 <sup>20</sup>	1	F: 1 (18 years)	L: 1	I: 1	Surgical	Improvement
Bekki et al. 2015 <sup>21</sup>	1	F: 1 (14 years)	T: 1	I: 1	Surgical	Improvement
Ge, Hao, Shan 2020 <sup>22</sup>	1	M: 1 (36 years)	L: 1	I: 1	Surgical	Improvement
Albulescu et al. 2016 <sup>23</sup>	1	U: 1 (45 years)	L: 1	I: 1	U	U
Cheng, Li, Lin 2012 <sup>5</sup>	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 <sup>24</sup>	1	F: 1 (23 years)	L: 1	I: 1	Non-Surgical	U
Hamidi and Foladi 2019 <sup>25</sup>	1	M: 1 (48 years)	L: 1	I: 1	Non-Surgical	U
Apostolopoulou 2021 <sup>26</sup>	1	F: 1 (5 years)	L: 1	I: 1	Surgical	Improvement
Kapsalakis 1964 <sup>27</sup>	2	F: 2 (6 years and 5 years)	L: 2	I: 2	Surgical	Improvement: 1/2
Vissarionov et al. 2018 <sup>28</sup>	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 <sup>29</sup>	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 <sup>30</sup>		months)				
Lersten, Duhon, Laker 2017 <sup>31</sup>	1	F: 1 (50 years)	L: 1	I: 1	Non-Surgical	U
Winter et al. 1974 <sup>32</sup>	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 <sup>33</sup>	1	F: 1 (55 years)	C: 1	II: 1	Non-Surgical	Improvement
Kim et al. 1994 <sup>34</sup>	5	M: 3, F: 2 Mean=14.8 years	L: 5	I: 5	Surgical: 5	Improvement: 4/5
Singh, Singh, Kumar 2015 <sup>35</sup>	1	F: 1 (3 months)	T: 1	I: 1	Surgical	U
Mamo, Batra, Steinig 2021 <sup>36</sup>	1	M: 1 (50 years)	T: 1	I: 1	Non-Surgical	U
McNeil, Jose, Rowland-Hill 2018 <sup>37</sup>	1	F: 1 (3 years)	L: 1	I: 1	U	U
Hader et al. 1999 <sup>38</sup>	1	F: 1 (16 years)	L: 1	I: 1	Surgical	U
Alimli et al. 2015 <sup>39</sup>	1	F: 1 (4 years)	T: 1	I: 1	Surgical	U
Huang et al. 2014 <sup>40</sup>	82	M: 17, F: 65 Median=6 years	T: 50 L: 32	I: 82	U	U
Khurram et al. 2021 <sup>41</sup>	1	M: 1 (38 years)	L: 1	I: 1	U	U
Shorey 1955 <sup>42</sup>	1	M: 1 (12 years)	L: 1	I: 1	Surgical	Improvement
Azimi and Mohammadi 2013 <sup>43</sup>	1	M: 1 (53 years)	L: 1	I: 1	Surgical	Improvement
Gavriliu et al. 2014 <sup>44</sup>	2	M: 1 (12 years), F: 1 (7 years)	T: 1 L: 1	I: 2	Surgical	Improvement: 1/2
Scotti et al. 1980 <sup>8</sup>	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 <sup>45</sup>	1	M: 1 (1.5 years)	L: 1	I: 1	Surgical	No improvement
English and Malthy 1967 <sup>46</sup>	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 <sup>47</sup>	1	F: 1 (5 years)	T: 1	I: 1	Surgical	Improvement
Gan et al. 2007 <sup>48</sup>	17	M: 8, F: 9 Mean=3.4 years	T: 5 L: 12	I: 17	Surgical: 17	Improvement: 5/17
Yamanaka et al. 2001 <sup>49</sup>	1	U: 1 (6 days)	L: 1	I: 1	Surgical	Improvement
Beyerl et al. 1985 <sup>50</sup>	1	M: 1 (34 years)	C: 1	I: 1	Surgical	Improvement
Sandhu, Gupta, Katha 2021 <sup>51</sup>	1	M: 1 (25 years)	T: 1	II: 1	Non-Surgical	Improvement
Chembolli 2015 <sup>52</sup>	1	F: 1 (16 years)	T: 1	I: 1	Surgical	Improvement
Elmaci et al. 2001 <sup>53</sup>	1	M: 1 (42	L:1	I: 1	Surgical	Improvement

		years)				
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Xu et al. 2023 <sup>54</sup>	1	F: 1 (17 years)	L: 1	I: 1	Surgical	Improvement
Zaleska-Dorobisz et al. 2010 <sup>55</sup>	1	F: 1 (78 years)	L: 1	I: 1	U	U
Sheehan, Sheehan, Lopes 2002 <sup>56</sup>	1	F: 1 (38 years)	T: 1	I: 1	Surgical	Improvement
Kanbur et al. 2004 <sup>57</sup>	1	M: 1 (12 years)	L: 1	I: 1	Non-Surgical	No improvement
Tubbs et al. 2004 <sup>58</sup>	1	F: 1 (18 years)	T: 1	I: 1	Surgical	No improvement
Shivapathasundram and Stoodley 2012 <sup>59</sup>	1	F: 1 (8 years)	T: 1	I: 1	Surgical	Improvement
Parmar et al. 2003 <sup>60</sup>	1	F: 1 (34 years)	L: 1	I: 1	Surgical	Improvement
Senkoylu et al. 2019 <sup>61</sup>	1	F: 1 (4 years)	L: 1	I: 1	Surgical	Improvement
Tsitsopoulos et al. 2006 <sup>62</sup>	1	F: 1 (44 years)	L: 1	I: 1	Non-Surgical	No improvement
Pettorini et al. 2007 <sup>63</sup>	1	M: 1 (2 years)	T: 1	I: 1	Surgical	Improvement
Lewandowski, Rachlin, Glazer 2004 <sup>64</sup>	1	F: 1 (44 years)	L: 1	I: 1	Surgical	Improvement
Ross et al. 1988 <sup>65</sup>	1	M: 1 (63 years)	S: 1	I: 1	Non-Surgical	No improvement
Porensky, Muro, Ganju 2007 <sup>66</sup>	1	F: 1 (54 years)	T: 1 L: 1	I: 2	Surgical	Improvement
Filippi et al. 2010 <sup>67</sup>	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 <sup>68</sup>	1	F: 1 (14 months)	T: 1	I: 1	Surgical	U
Shen et al. 2016 <sup>69</sup>	21 4	M: 61, F: 153 Mean=14.2 years	U	I: 73 II: 141	Surgical: 214	U
Sharma et al. 1997 <sup>70</sup>	1	M: 1 (15 years), F: 1 (9 years)	T: 1 L: 1	I: 2	Surgical: 2	U
Sgouros 2010 <sup>71</sup>	1	F: 1 (3 years)	L: 1	I: 1	Surgical	Improvement
Wenger, Hauswirth, Brodhage 2001 <sup>72</sup>	1	F: 1 (38 years)	L: 1	II: 1	Non-Surgical	Improvement
Kilickesmez, Barut, Tasdemiroglu 2004 <sup>73</sup>	1	F: 1 (7 years)	L: 1	I: 1	Surgical	Improvement
Kaminker et al. 2000 <sup>74</sup>	1	M: 1 (38 years)	L: 1	I: 1	Surgical	Improvement
Ak, Atalay, Gulsen 2014 <sup>75</sup>	1	F: 1 (10 years)	L: 1	I: 1	Surgical	Improvement
Ohwada, Okada, Hayashi 1989 <sup>76</sup>	1	M: 1 (29 years)	C: 1	I: 1	Surgical	Improvement
Morelli and Shalick 2011 <sup>77</sup>	1	F: 1 (29 years)	L: 1	I: 1	Surgical	No improvement
Macht et al. 2012 <sup>78</sup>	1	M: 1 (57 years)	L: 1	I: 1	Non-Surgical	U
Kanagaraju et al. 2016 <sup>79</sup>	1	F: 1 (15 years)	L: 1	I: 1	Non-Surgical	Improvement
Boussaandani et al. 2011 <sup>80</sup>	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 <sup>81</sup>						
Giordano et al. 2016 <sup>82</sup>	1	F: 1 (43 years)	T: 1	II: 1	U	U
Sharma et al. 2005 <sup>83</sup>	1	F: 1 (18 months)	L: 1	I: 1	Surgical	Improvement
Sedzimir, Roberts, Occleshaw 1973 <sup>84</sup>	1	M: 1 (22 months)	L: 1	I: 1	Surgical	Improvement
Callari and Arrigo 2009 <sup>85</sup>	2	F: 2 (80 and 59 years)	L: 2	U	Non-Surgical	Improvement: 2/2
Kramer, Dvorak, Curt 2009 <sup>86</sup>	1	F: 1 (54 years)	L: 1	I: 1	Surgical	No improvement
Roche and Vignaendra 2006 <sup>87</sup>	8	M: 3, F: 5 Mean=53.8 years	L: 7 S: 1	U	U	U
Uzumcugil et al. 2003 <sup>88</sup>	18	M: 2, F: 16 Mean=20 months	T: 9 L: 9	U	Surgical	U
Burnei et al. 2015 <sup>89</sup>	1	F: 1 (18 years)	L: 1	I: 1	Surgical	Improvement
Hung et al. 2010 <sup>90</sup>	1	F: 1 (2 days)	L: 1	I: 1	Surgical	U
Turgut and Doger 2008 <sup>91</sup>	1	M: 1 (1 day)	L: 1	I: 1	Surgical	U
Bale 1973 <sup>92</sup>	1	F: 1 (6 days)	T: 1	I: 1	Surgical	U
Korinth et al. 2004 <sup>93</sup>	1	M: 1 (2 years)	C: 1	I: 1	Surgical	No improvement
Mendez, Prieto, Lanciego 2009 <sup>94</sup>	1	F: 1 (88 years)	L: 1	I: 1	Non-Surgical	Improvement
Yamada, Mandybur, Thompson 1996 <sup>95</sup>	1	F: 1 (2 years)	L: 1	I: 1	Surgical	Improvement
Lourie and Bierny 1970 <sup>96</sup>	1	F: 1 (7 years)	T: 1	I: 1	Surgical	U
Ugarte, Gonzalez-Crussi, Sotelo-Avila 1980 <sup>97</sup>	2	F: 2 (1 day and 1 day)	T: 2	U	Surgical	Died: 2/2
Okada et al. 1986 <sup>98</sup>	1	M: 1 (19 years)	C: 1	I: 1	Non-Surgical	U
Azhar, Winter, Dunn 1996 <sup>99</sup>	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**

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