Research Article

A simple preoperative index for neurological deterioration in children with cervical and thoracic split cord malformation: development and temporal validation of the P(ND) score

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Abstract

Background: Children with split cord malformation (SCM) of the cervical and thoracic spine often present only after a neurological deficit has developed, and no validated tool estimates who will deteriorate before surgery. We developed a simple preoperative index, P(ND), from routine imaging and neurophysiology and tested it in a later cohort. Methods: Single-centre study of 42 children with cervical or thoracic SCM operated on in 2015–2026: a retrospective derivation cohort (n = 22, 2015–2021) and a prospective temporal validation cohort (n = 20, 2021–2026). Candidate predictors were tested against neurological status with Kendall's τ-b and combined into a weighted index. Discrimination was assessed by ROC analysis. Results: Spur asymmetry (τ-b = −0.72; $p < 0.001$), abnormal tibial somatosensory evoked potentials (τ-b = 0.60; $p = 0.01$) and the number of involved segments (τ-b = 0.45; $p = 0.02$) were retained, giving $P(ND) = 0.41 \cdot S + 0.34 \cdot E + 0.25 \cdot g(Seg)$. Across 42 children (32 with deterioration), the AUC was 0.924 (95% CI 0.843–1.000); at P(ND) ≥ 0.60, sensitivity was 93.8% and specificity 90.0%. Children treated under the P(ND)-guided protocol with ventral autologous duraplasty improved more often than historical controls (80.0% vs 40.9%; $p = 0.014$). Conclusion: P(ND) is a three-item index computed from standard MRI, CT and SSEP data that discriminated children with preoperative deterioration well in a single centre. External validation is needed before routine use.

Keywords: Split Cord Malformation; Diastematomyelia; Tethered Cord; Prognostic Model; Somatosensory Evoked Potentials; Children.

1. Introduction

Neural tube defects occur in about 4–5 per 10,000 births in Europe and remain more frequent in many low- and middle-income regions [1, 2]. Most of the literature concerns lumbosacral lesions. Dysraphism of the cervical and thoracic spine is rare, accounting for roughly 1–5% of cases, and has its own embryological and clinical features [3, 4].

Split cord malformation (SCM) is a closed form in which the cord is divided into two hemicords by a bony or fibrous septum [5]. Because external stigmata are often absent or subtle, children frequently come to surgery after a deficit has already appeared [6, 7]. In a recent series of 41 patients with type I SCM, more than half showed progression of congenital deficit or new deficit before the age of two years [8]. Early surgery is therefore widely recommended, yet the course is heterogeneous: some children deteriorate quickly while others remain stable for years [8–11].

Current guidance relies on clinical judgement and qualitative imaging [12, 16, 17]. Published series have not identified reliable preoperative predictors of deterioration, and outcome reporting in tethered cord surgery is inconsistent [8, 18]. Neurophysiological tests such as tibial somatosensory evoked potentials (SSEPs) can detect subclinical damage in closed dysraphism [14, 15], but they have not been combined with morphological findings into a single risk estimate.

We aimed to develop a simple preoperative index of neurological deterioration, P(ND), from data available in every routine work-up (MRI, CT and SSEP), and to test it unchanged in a later, independent cohort of children with cervical and thoracic SCM. Reporting follows the TRIPOD statement where applicable [22].

2. Materials and methods

2.1 Study design and patients

This single-centre retrospective–prospective cohort study was carried out in the paediatric neurosurgery department of the Republican Specialized Scientific-Practical Medical Center of Neurosurgery, Tashkent. It is part of a larger study of 102 children with cervical and thoracic spinal dysraphism operated on in 2015–2026; the present analysis includes the 42 children with SCM.

Inclusion criteria were age under 18 years, SCM of the cervical or thoracic spine confirmed by MRI and/or CT, and at least 6 months of postoperative follow-up. Exclusion criteria were purely lumbosacral lesions, incomplete records, severe associated malformations that could independently affect neurological status, and contraindications to surgery. The derivation cohort included 22 children operated on in 2015–2021, whose data were collected retrospectively. The validation cohort included 20 children operated on in 2021–2026, whose data were collected prospectively. No child contributed to both cohorts. The study was approved by the institutional ethics committee [committee name, approval number, date]. Written informed consent was obtained from parents or legal guardians.

2.2 Neurological assessment and outcome

Neurological status was graded with the Spina Bifida Neurological Scale (SBNS; motor 0–5, sensory 0–4, bladder/bowel 0–2; total 0–11, higher is better) [13]. Scales designed for adults (ASIA, JOA, ODI) were not used because they are unreliable in infants.

The primary outcome was preoperative neurological deterioration, defined as new onset or worsening of motor, sensory or sphincter deficit during the natural course of the disease before surgery. Postoperative deterioration was defined as worsening relative to the preoperative state.

2.3 Imaging and neurophysiology

All children underwent MRI of the whole spine (T1- and T2-weighted sagittal and axial images, coronal when required), under sedation in young children. CT (Philips MX16 EVO) was used to characterise the bony septum and vertebral anomalies. SCM was classified as type I or II according to Pang [5]. Spur symmetry was recorded as symmetric (midline) or asymmetric (lateralised), and the number of segments involved by the split was counted on MRI.

Electroneuromyography was performed on an INNOMED system. Tibial nerve (n. tibialis posterior) SSEPs were recorded and classified as normal for age or abnormal (prolonged latency or absent response).

2.4 Candidate predictors and index construction

Five candidate predictors available before surgery were tested in the derivation cohort: lesion level, presence of a bony spur, spur symmetry, number of involved segments, and tibial SSEP findings. Because the sample was small and SBNS is ordinal, associations with neurological status were assessed with Kendall's τ-b. Predictors with $p < 0.05$ were retained. Each retained predictor was scored on a simple scale (Table 1). Weights were set proportional to the absolute τ-b values and scaled to sum to 1.0 (0.72/1.77 = 0.41; 0.60/1.77 = 0.34; 0.45/1.77 = 0.25), giving:

$$P(ND) = 0.41 \cdot S + 0.34 \cdot E + 0.25 \cdot g(Seg)$$

Table 1. Scoring of P(ND) components

Component	Symbol	Score
Bony spur position	S	Symmetric = 0; asymmetric (lateralised) = 1
Tibial SSEP	E	Latency normal for age = 0; prolonged or no response = 1
Number of involved segments	g(Seg)	1 segment = 0; 2 segments = 0.5; ≥3 segments = 1

The index ranges from 0 to 1. Children were grouped as low (< 0.30), intermediate (0.30–0.59) or high risk (≥ 0.60). By construction, the high-risk category requires an asymmetric spur together with at least one other adverse feature; a single factor alone never reaches it. The formula and thresholds were fixed after derivation and applied unchanged to the validation cohort.

Surgical technique

In the derivation (historical) cohort, surgery consisted of spur removal without intradural ventral reconstruction, and neurophysiology was performed only before and after surgery. In the validation cohort, the timing and extent of surgery were planned with P(ND). Operations were performed under continuous intraoperative neuromonitoring (transcranial motor evoked potentials and direct stimulation) [19]. After laminectomy, the spur was thinned with a high-speed drill and removed, followed by intradural revision and untethering. The ventral dural defect was closed with a modified autologous rotation duraplasty: an elliptical flap of dorsal dura was rotated beneath the hemicords to the ventral defect and fixed with interrupted 6-0 polypropylene sutures, and the dorsal dura was then closed primarily and tested with a Valsalva manoeuvre.

Statistical analysis

Analyses were performed with IBM SPSS Statistics 27.0. Normality was tested with the Shapiro–Wilk test. Data are given as mean ± SD or median [Q1; Q3]. Groups were compared with Student's or Welch's t-test, the Mann–Whitney U test, ANOVA, χ^2 or Fisher's exact test as appropriate. Discrimination of P(ND) was assessed with the area under the ROC curve (AUC) and its 95% CI; sensitivity, specificity, predictive values and accuracy were calculated at the Youden-optimal threshold. A two-sided $p < 0.05$ was considered significant.

3. Results

3.1 Patient characteristics

Forty-two children with SCM were included: 22 in the derivation cohort and 20 in the validation cohort. Twenty-two children (52.4%) were younger than 1 year at surgery, 10 (23.8%) were 1–3 years old and 10 (23.8%) were older than 3 years. Type I SCM was found in 39 children (92.9%) and type II in 3 (7.1%). Tethering signs were present on MRI in all 42 children. The septum was located at Th7–Th12 in 29 children (69.0%), Th2–Th6 in 9 (21.4%), C5–Th1 in 2 (4.8%) and C1–C4 in 2 (4.8%). The cohorts did not differ in SCM type, septum morphometry or preoperative symptoms (Table 2).

Table 2. Baseline characteristics of the derivation and validation cohorts

Characteristic	Derivation, 2015–2021 (n = 22)	Validation, 2021–2026 (n = 20)	p
SCM type I / II, n	21 / 1	18 / 2	> 0.05
Septum height, mm (mean ± SD)	17.73 ± 1.45	17.40 ± 2.09	0.563
Septum thickness, mm	6.14 ± 2.23	6.55 ± 2.65	0.589
Spinal canal width, mm	16.05 ± 2.03	16.00 ± 1.86	0.940
Paresis or plegia, n (%)	18 (81.8)	17 (85.0)	> 0.05
Hypo- or anaesthesia, n (%)	21 (95.5)	19 (95.0)	> 0.05
Bladder/bowel dysfunction, n (%)	21 (95.5)	20 (100.0)	> 0.05
Clubfoot, n (%)	3 (13.6)	2 (10.0)	> 0.05
Trophic skin changes, n (%)	2 (9.1)	1 (5.0)	> 0.05

Preoperative SBNS fell with age at surgery: 10.32 ± 0.47 in children under 1 year, 9.70 ± 1.49 at 1–3 years and 8.20 ± 1.91 above 3 years (ANOVA $F = 10.46$; $p < 0.001$).

3.2 Predictor selection

In the derivation cohort, spur asymmetry showed the strongest association with neurological status, followed by SSEP findings and the number of involved segments (Table 3). Lesion level and presence of a bony spur were not associated with neurological status and were excluded.

Table 3. Association of candidate predictors with neurological status in the derivation cohort (n = 22)

Candidate predictor	Kendall's τ-b	p	Retained
Spur symmetry*	−0.72	< 0.001	Yes
Tibial SSEP	0.60	0.01	Yes
Number of involved segments	0.45	0.02	Yes
Lesion level	0.21	0.19	No
Presence of bony spur	0.18	0.28	No

*Negative sign: an asymmetric spur was associated with lower SBNS.

3.3 Performance of P(ND)

In the derivation cohort, P(ND) ≥ 0.60 identified all children with deterioration (sensitivity 100%) with a specificity of 80%. In the validation cohort, 2 children (10%) were classified as low risk, 7 (35%) as intermediate and 11 (55%) as high risk. Several high-risk children had minimal deficit at presentation, while MRI and SSEP showed persistent tethering; in these children surgery was offered on the basis of the predicted risk rather than symptoms alone.

Across all 42 children, 32 (76.2%) had preoperative deterioration and 10 (23.8%) were stable. The AUC was 0.924 (95% CI 0.843–1.000; p < 0.001). The Youden index peaked at 0.838 at P(ND) = 0.60 (Table 4, Figure 1).

Table 4. Classification by P(ND) at the 0.60 threshold, both cohorts combined (n = 42)

P(ND)	Deterioration	No deterioration	Total
≥ 0.60	30	1	31
< 0.60	2	9	11
Total	32	10	42

At this threshold, sensitivity was 93.8% (30/32), specificity 90.0% (9/10), positive predictive value 96.8% (30/31), negative predictive value 81.8% (9/11) and overall accuracy 92.9% (39/42).

3.4 Surgical outcomes

No deaths occurred. Follow-up ranged from 6 months to 9 years 7 months (median 3.5 years) in the full study population. Neurological improvement after surgery was more frequent in the P(ND)-guided cohort than in the historical cohort (16/20, 80.0% vs 9/22, 40.9%; Fisher's exact p = 0.014). Postoperative deterioration occurred in 1 (5.0%) and 2 (9.1%) children, respectively (Table 5). Two children had transient urinary retention that resolved with conservative treatment.

Table 5. Postoperative neurological outcomes in children with SCM

Outcome	Validation, P(ND)-guided (n = 20)	Derivation, historical (n = 22)	p*
Overall neurological improvement	16 (80.0%)	9 (40.9%)	0.014
Stable	3 (15.0%)	11 (50.0%)	—
Deterioration	1 (5.0%)	2 (9.1%)	—
Sensory improvement	10 (50.0%)	5 (22.7%)	0.107
Motor improvement	9 (45.0%)	4 (18.2%)	0.096
Bladder/bowel improvement	10 (50.0%)	7 (31.8%)	0.346
Persistent sensory deficit, long term	9 (45.0%)	16 (72.7%)	0.115
Persistent sphincter deficit, long term	10 (50.0%)	14 (63.6%)	0.533
Persistent motor deficit, long term	8 (40.0%)	14 (63.6%)	0.216

*Fisher's exact test.
Postoperative SBNS increased in all age groups. The between-group difference was significant only in infants (10.90 ± 0.30 vs 10.30 ± 0.30; $p < 0.001$), who also differed before surgery (10.60 ± 0.49 vs 10.08 ± 0.29; $p = 0.010$).

4. Discussion

We developed a three-item index that combines spur asymmetry, tibial SSEP findings and the extent of the split. In a single centre it separated children with and without preoperative deterioration well (AUC 0.924). All three components come from the standard work-up, so the index needs no additional tests and can be calculated at the bedside.
Each component has a plausible mechanism. A lateralised spur fixes the hemicords asymmetrically and concentrates traction and compression on one side, consistent with Pang's account of the clinical syndrome [6]. A prolonged or absent SSEP reflects loss of functional reserve and may precede clinical change; electrodiagnosis has detected neural damage even in asymptomatic infants with closed dysraphism [14, 15]. The number of involved segments indicates how far the malformation extends. The presence of a spur and the level of the lesion were not associated with deterioration, which suggests that the configuration of the septum matters more than its existence.
Our findings agree with the growing support for early surgery in type I SCM [8, 10, 11]. Narayanan and Rajshekhar found progression before the age of two in more than half of their patients but did not identify predictors [8]. P(ND) addresses this gap: it moves the question from "operate early" to "operate early in children at high risk". In our validation cohort, some children with minimal deficit were classified as high risk and operated on before the deficit deepened.
The P(ND)-guided protocol with ventral autologous duraplasty was associated with more frequent neurological improvement than historical treatment (80.0% vs 40.9%). This comparison cannot isolate the effect of the index. The two cohorts differ in era, intraoperative monitoring and dural reconstruction, and these factors may all contribute. Duraplasty has been linked to lower retethering rates in some, but not all, series [20, 21].

4.1 Limitations

This study has important limitations. First, the sample is small (42 children, 10 without deterioration), so confidence intervals are wide and the model may be overfitted; sample size calculations for prediction models suggest that larger cohorts are needed [23]. Second, weights were derived from univariable rank correlations, not from a multivariable regression, and the threshold was chosen on the data. Third, because the validation cohort contained few children without deterioration, discrimination was estimated in both cohorts combined; this estimate includes the derivation data and is therefore optimistic. Fourth, predictors and outcome were recorded before surgery in the same work-up, so the index describes current risk rather than a long-term forecast. Fifth, the historical control design is open to period effects. Finally, inter-observer reliability of spur symmetry and segment count was not assessed. External, multicentre validation is required.

5. Conclusion

$P(ND) = 0.41 \cdot S + 0.34 \cdot E + 0.25 \cdot g(Seg)$ is a simple preoperative index of neurological deterioration in children with cervical and thoracic SCM. It uses only routine MRI, CT and SSEP data and discriminated well in a single centre. A value of 0.60 or above, meaning an asymmetric spur plus at least one other adverse feature, may help identify children who benefit from early surgery. The index should complement, not replace, clinical judgement until it has been validated externally.

Declarations

Ethics approval and consent. Approved by [ethics committee], protocol No. [ ], [date]. Written informed consent was obtained from parents or guardians.
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Author contributions. J.M.A.: conception, data collection, analysis, drafting. M.M.A.: supervision, surgery, critical revision. [Others.]

Software registration. The P(ND) calculator is registered as software No. DGU 54904 (Uzbekistan).
Use of AI-assisted technology. A generative AI tool (Claude, Anthropic) was used to help draft and edit the English text from the authors' dissertation. The authors checked all content and data and take full responsibility for the manuscript.

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24. Figure legend
25. Figure 1. ROC curve of the P(ND) index for preoperative neurological deterioration in children with cervical and thoracic split cord malformation (n = 42). [Insert dissertation Figure 3.1 as a separate high-resolution file.]