# Surgical Management of Pediatric Nodular Lymphocyte Predominant Hodgkin's Lymphoma: Systematic Review and Case Report

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## Introduction

Nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL) is an uncommon subtype of Hodgkin lymphoma, representing about 5% of cases. It is defined by the presence of "popcorn" cells expressing CD20 and CD79a but lacking CD15 and CD30, features that distinguish it from classical Hodgkin lymphoma. Pediatric NLPHL most often affects males between 4 and 17 years of age, typically presenting with isolated cervical or axillary lymphadenopathy and favorable long-term survival rates exceeding 90%.

While conventional treatment with chemotherapy and radiotherapy is effective, these modalities are associated with significant late toxicities, including impaired growth, infertility, and secondary malignancies. In recent years, surgical resection alone has emerged as a potential management strategy for selected early-stage, low-risk pediatric patients, paralleling treatment approaches in other childhood malignancies such as Wilms tumor and neuroblastoma. This systematic review, combined with a representative case, evaluates the role of surgery as a primary treatment for pediatric NLPHL and its potential to reduce treatment-related morbidity while maintaining excellent survival outcomes.

## Methods

A systematic review was conducted using PubMed to identify studies published between 2003 and 2024 that examined surgical management of pediatric nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL). Search terms included "surgical treatment" and "pediatric NLPHL." Studies were included if they focused on pediatric patients and surgical resection as a primary therapy, while those emphasizing chemotherapy, radiation, mixed pathology, or adult populations were excluded. Quality assessment of eligible studies was performed using the Downs and Black risk of bias tool, and methods adhered to PRISMA guidelines.

In addition to the literature review, we present the case of an 11-year-old male with unilateral cervical NLPHL who underwent modified radical neck dissection. Clinical, pathologic, and imaging findings were reviewed, along with disease course and treatment outcomes. Together, these data provide a framework for evaluating the role of surgery as a frontline therapy for pediatric NLPHL.

## Results

Four pediatric NLPHL studies met inclusion criteria, representing patients aged 4–17 years treated primarily with surgical excision. Across studies, surgical management demonstrated excellent overall survival (100%) and variable event-free survival (57–77%), with outcomes strongly influenced by the extent of resection. Complete nodal excision was consistently associated with durable remission, while incomplete resections carried higher relapse rates that were usually salvageable with chemotherapy.

| ARTICLE CITATION | NUMBER OF PATIENTS | TREATMENT TYPE | INDICATION FOR TREATMENT | OUTCOME AFTER TREATMENT | REMISSION STATUS | OTHER INFORMATION |
|---|---|---|---|---|---|---|
| Appel et al., 2016 | 178 | Surgery alone for resected single node in stage 1A patients, AV-PC chemotherapy with/without RT for unresected stage I/II patients or those with less than a complete response to surgery. | Stage IA/IIA NLPHL, surgery for resected nodes, AV-PC chemotherapyfor unresected nodes or relapses | For surgery alone: 52 patients had a 5-year event-free survival (EFS) 77%, overall survival (OS) 100%. 9 of the patients had to undergo AV-PC. For AV-PC chemotherapywith/without RT: EFS 88.8%, OS 100%. Recurrences were treatable. | 75% of patients achieved complete remission after surgery alone; recurrence mostly treatable with chemotherapy. | Patients were followed up for 5 years; relapses in surgery-only group occurred within 2 years post-treatment. |
| Pellegrino et al., 2003 | 27 | Surgery alone for resected nodes or combined therapy (chemotherapy/radiotherapy) for residual disease or advanged stages. | Stage I (n=22), Stage II (n=2), Stage III (n=3) NLPHL | 23 of 27 patients achieved CR; OS 100%; EFS 69% ± 10%. For SA alone group, EFS was 42% ± 16%; for CT group, EFS was 90% ± 8.6% | Higher relapse rates in surgery-only group for those with residual disease, but remission was achieved overall with combined therapy. | Relapse was localized, mostly mangeable with additional treatments; OS remained high. |
| Mauz-Korhölz et al., 2007 | 58 | Surgery alone for early-stage, limited disease (stage IA/IIA). | Early-stage, limited disease, mainly stage IA/IIA. | For surgery alone: Progression-free survival (PFS) 57%, OS 100%. Relapse occurred in 7 (12.07%) patients with residual disease. Patients with complete resection had better outcomes. | Relapses primarily occurred in patients with residual disease, long-term remission achieved in others. | Residual disease was a significant factor in relapse rates. Long-term follow-up showed favorable OS. |
| Bessen et al., 2021 | 1 | Surgery alone initially, followed by chemotherapy after disease recurrence (4 year old patient) | Localized disease without systemic symptoms, reucrrence within 2 months after surgery. | Recurrence occurred 2 months after surgery, followed by chemotherapy treatment. | Required chemotherapy after recurrence, remission not sustained after surgery alone. | Short interval to recurrence suggests a more aggressive course requiring follow-up chemotherapy |

## Case Report

An 11-year-old male presented in July 2024 with a three-week history of progressive left neck swelling without systemic symptoms such as fever, weight loss, or night sweats. Laboratory workup, including CBC, CMP, LDH, and ESR, was within normal limits. Contrast-enhanced CT of the neck revealed multifocal unilateral cervical adenopathy, the largest node measuring 4.8 cm, without mediastinal or abdominal involvement. PET-CT confirmed Stage IA disease, with FDG uptake localized to the left cervical chain. Core biopsy demonstrated nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL), characterized by CD20+, PAX-5+, and BCL-6+ "popcorn cells," with CD15 and CD30 negativity.

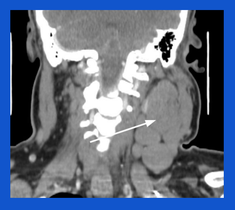


*Figure 1A.*

*Preoperative coronal CT demonstrating bulky left cervical lymphadenopathy (arrow), measuring up to 4.8 cm*

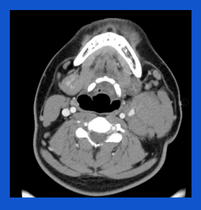


*Figure 1B.*

*Axial CT image showing homogeneous, enlarged lymph nodes at cervical level II*

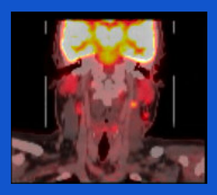


*Figure 1C.*

*Coronal PET-CT confirming isolated metabolic activity in the cervical nodal chain*

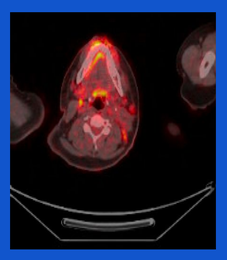


*Figure 1D.*

*Preoperative axial PET-CT showing FDG uptake localized to the left neck mass*

In August 2024, the patient underwent a modified radical neck dissection of levels II–V. Intraoperatively, innumerable firm lymph nodes were noted. Pathology revealed multiple nodes replaced by NLPHL, with classic morphologic features and immunophenotype confirming the diagnosis. Initially, postoperative PET demonstrated remission, with only mild uptake thought to be reactive. The patient was monitored closely with surveillance imaging.

By February 2025, repeat PET-CT showed increased size and number of cervical, supradiaphragmatic, and infradiaphragmatic lymph nodes with a Deauville score of 5, representing recurrent Stage IIIA disease. Multidisciplinary tumor board consensus recommended systemic therapy. A central venous port was placed, and the patient began R-CHOP chemotherapy. He completed four cycles by May 2025.

Post-treatment PET-CT demonstrated complete metabolic response with no abnormal FDG uptake. At nine months from initial presentation, the patient remains in remission with no evidence of disease (NED). This case illustrates both the potential for surgery as an initial management strategy in pediatric NLPHL and the importance of vigilant surveillance, as unexpected recurrence may necessitate systemic therapy.

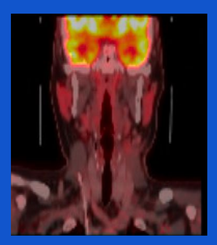


*Figure 1E.*

*Postoperative coronal PET-CT (March 2025) demonstrating complete metabolic resolution*

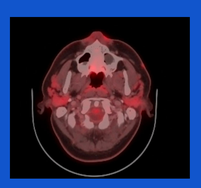
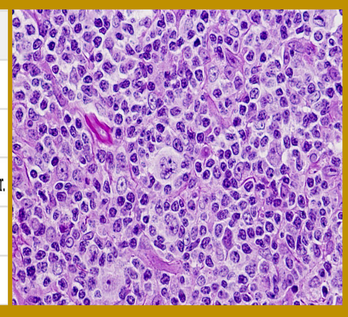


*Figure 1F.*

*Postoperative coronal PET-CT showing absence of hypermetabolic activity in the cervical and supradiaphragmatic regions*

| Deauville Score (Visual Score) | Criteria |
|---|---|
| 1 | No uptake. |
| 2 | Uptake ≤ mediastinal blood pool. |
| 3 | Uptake > mediastinal blood pool and ≤ normal liver. |
| 4 | Moderately increased uptake > normal liver. |
| 5 | Markedly increased uptake > normal liver. |



## Discussion

Our findings highlight surgical resection as a feasible frontline option for carefully selected pediatric NLPHL patients. Across four studies, complete excision was associated with event-free survival rates of 57–77% and overall survival of 100%, while incomplete resections carried higher recurrence risk but were typically salvageable with chemotherapy. These results mirror treatment strategies in other pediatric malignancies, such as Wilms tumor and low-risk neuroblastoma, where surgery alone has successfully minimized long-term toxicity.

The presented case reinforces both the promise and limitations of this approach. Although initial staging suggested suitability for surgery alone, high nodal burden and later recurrence necessitated systemic therapy. This underscores the importance of precise patient selection and close postoperative monitoring. Current evidence remains limited by small sample sizes and retrospective designs, emphasizing the need for prospective, multicenter trials to refine risk-adapted surgical protocols and to assess long-term quality of life outcomes.

## Conclusion

Surgical resection has emerged as a promising frontline strategy for pediatric patients with localized, low-risk nodular lymphocyte-predominant Hodgkin lymphoma (NLPHL). Across the reviewed studies, complete resection was consistently associated with excellent outcomes, achieving event-free survival rates of up to 77% and overall survival of 100%, while avoiding the long-term toxicities of chemotherapy and radiotherapy. These findings support surgery as a viable option in carefully selected cases, especially when complete excision is possible.

Our case illustrates both the potential and the limitations of this approach. Although the patient initially appeared to be a candidate for surgical management alone, a higher-than-expected tumor burden and subsequent recurrence required systemic therapy. This highlights the importance of accurate staging, careful patient selection, and close follow-up in order to maximize outcomes.

Future prospective, multicenter studies are essential to refine criteria for surgery-only management, establish standardized follow-up protocols, and evaluate long-term quality of life outcomes. By advancing risk-adapted approaches, clinicians may reduce overtreatment while preserving excellent survival for children with this rare lymphoma subtype.

References: