CASE SERIES

Functional outcomes of internal hemipelvectomy in paediatric patients with malignant bone tumours: a monocentric case series

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A case series was conducted between January 2010 and January 2018, which included 17 paediatric patients under the age of 18 years (mean age 13.4 ±3.53), who underwent internal hemipelvectomy. Functional outcomes were assessed based on gait and categorised as either normal or abnormal. The most common diagnosis was Ewings Sarcoma in 14 (82.3%) patients. With regards to reconstruction, acetabular reconstruction was necessary in 1(5.9%) patient, and soft tissue coverage with flaps was required in 5 (29.4%) cases. Nerve injury was the most common complication noted, i.e. in 5(29.4%) cases. The disease-free survival rate was 12 (70.6%) patients (70.6%) at a median time of 72 months, while the overall mortality rate was 5(29.4%) patients with metastatic disease being the leading cause. Hence, it was concluded that internal hemipelvectomy is a viable surgical option in the paediatric population particularly when negative margins can be achieved without compromising the neurovascular structures.

Keywords: Tumour, Paediatric, Internal hemipelvectomy.

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Introduction

Paediatric pelvic tumours are a rare occurrence, representing 5-15% of all bone tumours, with Ewing Sarcoma and Osteosarcoma being the most prevalent types.[1] Managing these tumours is challenging due to their large size, complicated anatomical location, and potential involvement of nearby neurovascular, genitourinary, and gastrointestinal systems.[2, 3] The deep location of these tumours within the pelvis often leads to delayed diagnosis, as patients typically present late with nonspecific signs and symptoms.[4]

The primary surgical approach for paediatric pelvic tumours used to be external hemipelvectomy; however,[2,4] with recent advancements in treatment, internal hemipelvectomy, has emerged as a viable option.[3] This approach is suitable for cases where tumours are smaller and confined to a hemipelvis without the involvement of the thigh and ilio-femoral bundle.[3] However, it is important to note that this procedure is not without complications. Wound infections, neurovascular injuries, implant failures, and high local recurrence rates leading to revisions or amputations are among the potential complications associated with internal hemipelvectomy.[5]

Internal hemipelvectomy has demonstrated effectiveness in the adult population, but there is a paucity of data on its outcomes in the paediatric age group due to the rarity of pelvic tumours in children.[6] Considering the longer life expectancy of children and having the potential to enhance their quality of life, limb salvage procedures hold particular significance in this population. This study aims to share our experience with this procedure in children treated at a tertiary care centre.

Case Series

For this retrospective case series conducted at Aga Khan University Hospital - a tertiary care centre in Karachi, Pakistan, 17 paediatric patients (age < 18 years) who

Table-2: Patient characteristics and outcomes

	Frequency (n)	Percentage (%)	SD	Range	Mean
Age (years)	17		3.53	17-May	13.4
Follow up	17		29.6	Aug-96	34.4
Gender					
Male	12	71			
Female	5	29			
Biopsy:					
External Source	10	58			
Within Hospital	7	42			
Diagnosis					
Ewings	14	82.3			
Osteosarcoma	2	11.7			
Synovial Sarcoma	1	5.9			
Types of Resections					
Type I	4	23.5			
Type III	1	5.9			
Type IV	1	5.9			

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Type I + II	2	11.8
Type I, II+ III	2	11.8
Type II + III	4	23.5
Type I + IV	1	5.9
Type I, II + IV	2	11.8
Acetabular Reconstruction		
Free suspension	11	64.7
Acetabular reconstruction	1	5.9
No reconstruction	5	29.4
Coverage With Flaps		
Yes	5	29.4
No	12	70.6
Margins status		
Positive Margins	1	5.9
Negative Margins	16	94.1
Complications	11	64.7
Infection	4	23.5
Nerve injury	5	29.4
Flap Necrosis	1	5.9
Local Recurrence	1	5.9
Cause of Expiry	9	52.9
Metastasis	4	23.5
Local recurrence	1	5.9
Chemotherapy induced	1	5.9
Non oncological reasons	3	17.6
Functional Outcome		
Ambulation with Normal Gait	10	58.8
Ambulation with Abnormal Gait	7	41.2

underwent internal hemipelvectomy between January 2010 and January 2018 were selected from the hospital's institutional tumour registry (Table 1). Approval was obtained from the head of the orthopaedic section and consent was taken from the parents of all patients for using the data anonymously. Tumours were classified by using the Enneking Staging System.[7] and the levels of resections were classified according to the Enneking and Dunnham Classification.8 (Figure 1).

Patients' files were reviewed to assess any complications and their management. Functional outcomes were evaluated based on gait and categorised as either normal or abnormal based on the surgeon's assessment. All surgeries were performed by the same musculoskeletal oncology surgeon who has specialised fellowship training. In cases requiring acetabular resection, reconstruction involved hip transposition, where the femoral head was repositioned to the proximal osteotomy level. A joint capsule was created around the head using the remaining iliopsoas and gluteus muscles, and if necessary, additional augmentation was performed using screws or bone anchors. For pelvic resections involving the pubic and ischial bone, soft tissue and joint capsule approximation were sufficient.

Table-1: Details of all included patients

Case No.	Location where patient presented	Age at presentation	Date of presentation	Gender	Diagnosis
1	AKUH*, Karachi, Pakistan	17	10/11/2005	Male	Osteosarcoma
2	AKUH*, Karachi, Pakistan	12	25/03/2010	Female	Ewings Sarcoma
3	AKUH*, Karachi, Pakistan	14	29/09/2011	Male	Ewings Sarcoma
4	AKUH*, Karachi, Pakistan	5	28/06/2012	Male	Ewings Sarcoma
5	AKUH*, Karachi, Pakistan	15	26/07/2012	Male	Ewings Sarcoma
6	AKUH*, Karachi, Pakistan	8	22/11/2012	Male	Ewings Sarcoma
7	AKUH*, Karachi, Pakistan	13	28/03/2013	Male	Ewings Sarcoma
8	AKUH*, Karachi, Pakistan	16	20/02/2014	Male	Osteosarcoma
9	AKUH*, Karachi, Pakistan	14	15/05/2014	Female	Ewings Sarcoma
10	AKUH*, Karachi, Pakistan	16	27/11/2014	Female	Ewings Sarcoma
11	AKUH*, Karachi, Pakistan	16	19/03/2015	Male	Ewings Sarcoma
12	AKUH*, Karachi, Pakistan	14	18/05/2015	Female	Ewings Sarcoma
13	AKUH*, Karachi, Pakistan	17	03/12/2015	Male	Synovial Sarcoma
14	AKUH*, Karachi, Pakistan	16	17/02/2017	Female	Ewings Sarcoma
15	AKUH*, Karachi, Pakistan	14	05/07/2017	Male	Ewings Sarcoma
16	AKUH*, Karachi, Pakistan	13	14/12/2017	Male	Ewings Sarcoma
17	AKUH*, Karachi, Pakistan	7	10/01/2018	Male	Ewings Sarcoma

*AKUH – Aga Khan University Hospital

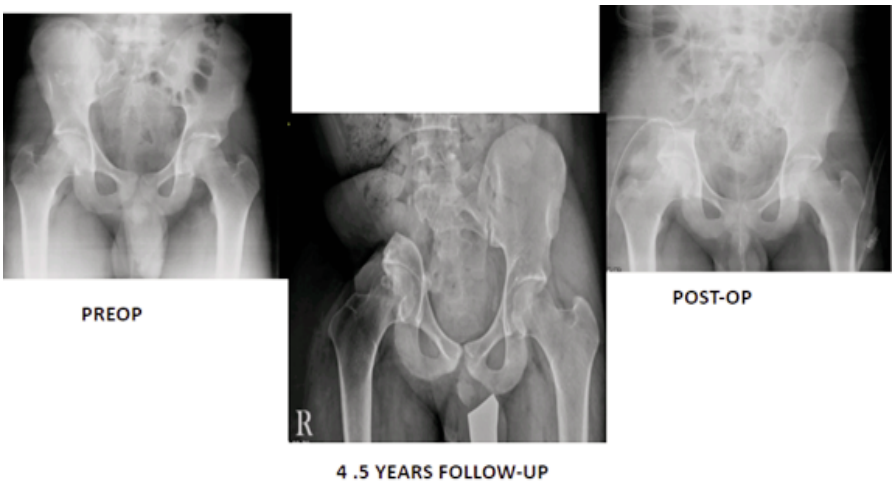

Figure-1: 16-year-old male, Diagnosis Synovial Sarcoma, Type 1 Internal Hemipelvectomy.

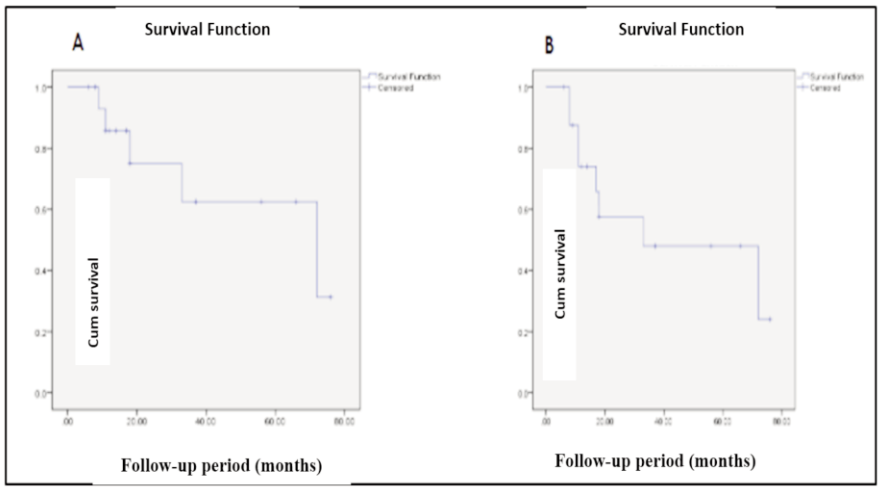

Figure-2: Showing Kaplan Meier survival analysis: A: The disease free survival, B: Overall Survival rate.

The mean age of the patients was 13.4±3.53 years and the mean duration of follow-up was 34.4±29.6 months (Table 2). Histological analysis revealed that 14 cases (82.3%) were diagnosed as Ewing's sarcoma, followed by 2 cases (11.7%) of osteosarcoma, and 1 case 5.9% of synovial sarcoma. Various levels of internal hemipelvectomies were performed, with type I and type II+III being the most common (Table 2). Acetabular reconstruction was necessary in only one (5.9%) patient, while 5 (29.4%) patients did not require any reconstruction. In 11 (64.7%) patients, the limb was managed with free suspension. Soft tissue coverage using flaps was required in 5 (29.4%) cases, and the negative margin rate was seen in 16 (94.1%) patients.

Complications were noted in 11 (64.7%) patients, with nerve injury being the most frequent in 5 (29.4%) cases, followed by local wound infection in 4 (23.5%) cases, and flap necrosis in 1 (5.9%) patient. One case of local tumour recurrence was also observed. Mortality was attributed to metastatic disease in 4(23.5%) patients and other reasons in 3(17.6%) patients. Additionally, local recurrence and chemotherapy-induced complications were the cause of death in two patients each. The disease-free survival was noted in 12 (70.6%) patients with a median follow-up time of 72 months (Inter-quartile range 20-80 months), while the overall survival was noted in 9 (52.9%) patients with a median follow-up time of 33 months (Inter-quartile range 18-72 months) (Figure 2).

Discussion

Pelvic tumours in paediatric populations tend to present late and pose a great challenge with regards to diagnosis and management.[2,5] When limb salvage is opted for with internal hemipelvectomy, a standard utilitarian incision is commonly given, as was utilised in all the patients in this study, with specific modifications tailored to ensure wide-margin excision. This approach allows for a broader exposure, enabling the complete removal of the tumour while avoiding contamination of surrounding tissues.[9] The enhanced exposure played a crucial role in achieving a high negative margin rate of 93.7% in the study population.

With regards to reconstruction, potential options include lumbopelvic fusion with pedicle screw instrumentation, specially where sacrum is involved. For acetabular involvement, a modular hemipelvis system may be utilised, connected to lumbopelvic fixation proximally and a mega prosthesis distally for hip reconstruction if needed. More conservative approaches involve iliofemoral fusions, suspension techniques, or a flail limb, which was the case for 5(62.8%) of the patients in this study.[6, 10, 11] While most patients do not need soft tissue coverage, 31.3% of the study patients required coverage with a flap, for which plastic surgery consultation was sought.

In this study, complications were observed in 11 patients, with the most frequent being post-operative neurological deficits and wound infections, as also reported in international literature.[6] These complications were primarily attributed to the extensive nature of soft tissue dissections and resection required for the procedure. Similar to the findings in other studies, infection rates were higher in patients who underwent reconstruction compared to those who did not require reconstruction.[12] Performing internal hemipelvectomy in the paediatric population is considerably challenging as the internal organs and limb structures in children are smaller and more delicate compared to adults, making them more susceptible to iatrogenic injury during the surgical procedure.[2]

Mortality was recorded in nine patients, with four of them succumbing to metastatic disease, which emerged as the primary cause. Despite these challenges, the disease-free survival rate was 70.6% over a median time period of 72 months. Internationally, five-year survival is reported to be around 50%.[6] Overall, the outcomes of patients who underwent internal hemipelvectomy in the current study were favourable, with limited complications and acceptable functional outcomes.

Internal hemipelvectomy is not a commonly performed procedure in the paediatric population and specialised training through multiple pelvic and acetabular courses is crucial for improved outcomes. Additionally, adopting a multidisciplinary approach is vital, which is often lacking in developing countries. There is scarcity of literature on internal hemipelvectomy in the paediatric population, while no prior data is available from our country. Therefore, this study offers valuable evidence regarding the use of this specific procedure in paediatric patients, even in resource-constrained settings with limited opportunities for specialised training compared to developed countries.

However, it is important to acknowledge the limitations of the study. The sample size and descriptive design of the study prevents us from drawing definitive conclusions. Furthermore, all surgeries were performed by a single surgeon, which limits the generalisability of the results. Due to the retrospective design of the study functional outcome scores could not be calculated. Therefore, further prospective studies with larger sample sizes are necessary to provide a more robust consensus on the outcomes and potential benefits of this surgical approach for paediatric pelvic tumours.

Conclusion

Based on the results of the current study, it can be said that internal hemipelvectomy is a feasible option even in paediatric population and should be considered when negative margins can be obtained, and tumour resection does not compromise neurovascular structures.

Ethics approval and consent to participate: This study was conducted as per relevant guidelines and regulations or Declaration of Helsinki.

Written informed consent was obtained from all subjects and/or their legal guardian(s) in ethical approval. The waiver for approval of study protocol was granted by Aga Khan University hospital ethics review committee (ERC). ERC reference number is 2020-0723-8710.)

Consent to publication: An informed consent from all subjects and/or their legal guardian(s) was taken for publication of identifying information/images in an online open-access publication anonomously.

Availability of data and materials: The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

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FAJ: Concept, design, data collection, analysis and writing.
JS: Data analysis and writing.
MA: Data analysis, results and writing.
MU: Concept, design, writing and revision.