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A retrospective study on the incidence and outcomes of hepatoblastoma in relation to prognostic indicators at Universitas Academic Hospital, Bloemfontein, South Africa (2000–2022)

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Background: Hepatoblastoma (HB) is the most common solid liver cancer in childhood. Globally, survival has significantly improved with advanced treatment modalities. However, survival is poor in low-middle-income countries (LMICs) due to advanced-stage presentation.

Aim: The aim was to evaluate the incidence and outcomes of children diagnosed with HB in relation to prognostic indicators which included age, sex, PRE-Treatment EXTent (PRETEXT) staging, metastasis and surgical intervention. Outcomes included survival and mortality.

Setting: The research was conducted at the Pediatric Oncology Unit in Universitas Academic Hospital in Bloemfontein, South Africa, which serves as a tertiary referal centre for patients from the Northern Cape, Free State and Lesotho.

Methods: We retrospectively analysed medical records of the children with HB diagnosed and treated from 2000 till 2022 at Universitas Academic Hospital Paediatric Oncology Unit.

Results: Twenty patients were included with an incidence of 1.2% and a 95% confidence interval (CI) of 0.8%–1.8%. Median age was 2 years old with interquartile range (IQR) of 1 year – 2.97 years. There was a male: female ratio of 1.9:1. All children presented with abdominal distension and elevated alpha fetoprotein (AFP) levels. More than half (55%; $n = 11$) had PRETEXT IV and 45% ($n = 9$) had lung metastasis at presentation. Surgical resection was done in 50% ($n = 10$) and none had liver transplant. The 1 year overall survival (OS) was 55% ($n = 11$), 2-year OS was 20% ($n = 4$), and 5 year OS was 10% ($n = 2$). Higher mortality was noted in children above 3 years old ($p = 0.5304$), male patients ($p = 0.1011$), distant metastasis ($p = 0.5913$), PRETEXT stage IV ($p = 0.2621$), high risk HB ($p = 0.117$) and children who did not get surgical resection ($p = 0.0867$).

Conclusion: Survival was negatively impacted by older age, male gender, advanced stage of disease and paucity of surgical intervention.

Contribution: This study highlights the factors affecting the survival of children with this rare disease and highlights measures to improve same.

Keywords: hepatoblastoma; pre-treatment extent of disease; lung metastasis; surgical resection; liver transplantation.

Introduction

Hepatoblastoma (HB) is the commonest primary liver tumour in the paediatric population.[1] It is, however, rare and represents 0.5%–2% of all childhood cancers.[1,2] The incidence of HB is approximately 1.5 per million population and has steadily increased by about 5% each year.[1,3,4,5] Hepatoblastoma is usually seen in children younger than 5 years of age.[4,6] Presenting symptoms include abdominal swelling and other non-specific symptoms like loss of weight, malaise and rarely, tumour rupture.[7,8,9]

A biopsy is done to confirm the histology if there is diagnostic uncertainty, and treatment is guided by staging and risk stratification.[10] Hepatoblastoma is classified as standard risk (SR–HB) when the tumour is confined to the liver, PRE-Treatment EXTent (PRETEXT) I, II, III without annotation factors, no metastasis and alpha fetoprotein (AFP) > 100 ng/mL and high risk (HR–HB) when metastasis has occurred, PRETEXT IV or any stage with annotation factors, tumour rupture and AFP < 100 ng/mL.[11]

With cisplatin-based chemotherapy, surgical intervention like tumour resection and liver transplantation (LT), children with SR–HB have good outcomes with survival up to 90%.[12,13] However, outcomes remained poor in children with HR–HB until the emergence of LT and survival improved to > 50%.[12,13] Disease advancement with relapsed or refractory HB also impacted negatively on survival, and some new modalities like thermal ablation have been employed.[14]

Surgical resection of the tumour is imperative for long-term cure.[8,15] Unfortunately, about 60%–70% of children present with an unresectable tumour due to great tumour bulk and spread to major blood vessels. No surgical intervention or incomplete removal of the tumour is associated with higher mortality.[11]

Late presentation and diagnosis with advanced disease, limited access to healthcare, unavailability of LT services, treatment toxicity and treatment abandonment negatively impact the survival of children with HB in low-middle-income countries (LMICs).[12] Malnutrition is still a huge burden in LMICs and adversely impacts outcome.[16] Hepatoblastoma is a rare tumour, and there are few studies done to scrutinise outcomes especially in LMICs. Hence, this study aims to analyse the incidence of HB and the outcomes of the children diagnosed with HB and managed at the Paediatric Oncology Unit of Universitas Academic Hospital (UAH) over a period of 23 years and compare our survival with other parts of the world.

Methods

A retrospective descriptive cohort study was conducted at the Paediatric Oncology Unit at Universitas Academic Hospital that included patients admitted and diagnosed with HB during the study period from January 2000 to December 2022. The incidence was calculated per number of admissions to the unit.

Annually, on average, 90 children are admitted to the unit, and of these about one case of HB is diagnosed per year. Based on this, we anticipated a sample size of approximately 22 patients – 30 patients.

Inclusion criteria

Included all children admitted to the unit as well as those diagnosed with HB within the study period.

Exclusion criteria

Missing medical records and children diagnosed outside the study period.

Ethical considerations

Ethics approval was granted by the Health Sciences Research Ethics Committee (HSREC) at the University of the Free State (Ethics number: UFS – HSD 2023/0217/2507), and permission to conduct the study was also obtained from the Free State Department of Health. Consent from participants was not required as it was a retrospective study. There was no contact between the researchers and the patients or their caregivers.

A data collection form was created on Redcap® with password protection and was used for data collection. Each participant was assigned a unique numerical identifier, thereby anonymity was maintained. No personal information was revealed to the public.

Twenty patients met the inclusion criteria. We audited the patient files and electronic records, and data were collected to include the following demographics: (1) age and sex, (2) constitutional genetic abnormalities, (3) malformations, (4) clinical presentation, (5) investigations done including full blood count, (6) platelet count, (7) AFP and liver function test, (8) histology and staging, and (9) management and outcomes namely survival and mortality. Management included medical treatment in the form of chemotherapy and surgical interventions which included surgical resection of the tumour. A pilot study was done with five participants to test the data collection tool, and some modifications were made like the exclusion of neonatal risk factors and maternal risk factors, such as smoking as there was scanty data on that. The results obtained from the pilot study were included in the final analysis. The collected data were anonymised before sending them to the biostatistician for statistical analysis.

Analysis

Descriptive statistics, namely median and percentiles, as the distribution of numerical data was skewed, and frequencies and percentages for categorical data were calculated. The incidence of HB was calculated and described using 95% confidence interval (CI) for the incidence. Associations between prognostic indicators and mortality were calculated and described using Fisher's exact test for sparse categorical data. The analysis was done by the Department of Biostatistics, University of the Free State (UFS). The data analysis for this study was generated using SAS software. Copyright, SAS Institute Inc. SAS and all other SAS Institute Inc. product or service names are registered trademarks or trademarks of SAS Institute Inc. (Cary, North Carolina, United States [US]). A p-value of < 0.05 was defined as significant.

Results

Twenty children who were diagnosed with HB were included in the study. Five patients whose medical records could not be found and one patient who was diagnosed in 1988 were excluded.

Incidence

The number of children diagnosed with HB was 25 patients of the total 2076 children admitted with cancer within the study period, with an incidence of 1.2% and a 95% CI of 0.8% – 1.8%.

Patient characteristics

The majority of the patients, 60% (*n* = 12), were referred from within the Free State province, 30% (*n* = 6) were from the Northern Cape province and 10% (*n* = 2) from Lesotho.

The median time from onset of symptoms to presentation at UAH was 2.7 weeks with interquartile range (IQR) of 1.3 weeks – 8 weeks, and the longest time noted before presentation was after 10 months of symptoms. Other clinical characteristics, investigations and results are presented in Table 1.

Investigations

In some children, an initial abdominal sonar was done to aid diagnosis. All the children had a computed tomography (CT) scan documented which was the most utilised radiological modality for staging. There was overlap with radiological investigations with most children having at least two modalities.

Biopsy was done in most of the patients and five did not have a documented biopsy. Rationale for non-performance of biopsy was not stated in the medical records (Table 1).

Histology showed the majority, 33% (*n* = 5), to be foetal and embryonal type, followed by mixed epithelial 27% (*n* = 4). Purely foetal was 13% (*n* = 2), and purely embryonal was 7% (*n* = 1). Thirteen per cent (*n* =2) showed foetal and mixed epithelial histology, and 7% (*n* = 1) showed macrotrabecular pattern.

Staging

Majority (*n* = 12; 60%) of the patients (with PRETEXT IV or metastasis) were classified as high risk at diagnosis and 35% (*n* = 7) as standard risk (PRETEXT I, II, III and no metastasis) (Table 2).

Treatment

One child did not receive chemotherapy because the patient died 4 days after diagnosis before commencement of chemotherapy. For relapses and refractory HB, 5-fluorouracil and irinotecan were used. The number of chemotherapy courses given was a median of 6.00 with an IQR of 2.5 courses to 8 courses. Eight children had more than six courses of chemotherapy due to unfavourable response or disease progression. Surgical resection was possible in only 50% (*n* = 10) of the patients, and most patients were too unstable for surgery with advanced PRETEXT staging and high-risk stratification (Table 2).

Relationship between risk stratification and response to chemotherapy

Of the 20 children, 35% (*n* = 7) achieved remission, six (85%) of which were classified as standard risk. Unfortunately, 42% (*n* = 3) of them relapsed. Of the three patients who relapsed, the period to relapse was after 5 months in one patient and 12 months in the other two patients. In contrast, for the high-risk patients, which were 60% (*n* = 12), only one achieved remission and the majority, 91% (*n* = 11) had disease progression with poor or no response to chemotherapy (Figure 1).

Survival

At the end of the study period, 80% (*n* = 16) of the children had been documented to have died. After diagnosis, the longest survival duration was 3 years and 7 months and the shortest was 4 days (IQR of 0.22 months–1.02 years). Two patients (10%) had abandoned treatment. Both abandoned treatment after eight courses of chemotherapy at 13 months and 14 months post starting treatment, respectively. The remaining two patients (10%) were documented to be alive and still being reviewed.

For all the children, 1 year overall survival (OS) was 55% (*n* = 11), 2 year OS was 20% (*n* = 4), 2 year event free survival was 10% (*n* = 2), 5 year OS was 10% (*n* = 2) and 5 year event free survival was 10% (*n* = 2) (Table 2).

Survival in relation to prognostic indicators

For the standard risk patients, 1 year OS was 85% as compared to the high-risk patients which was 33% ($p = 0.147$); 2 year OS was 42% for standard risk patients and 8% for high-risk patients ($p = 0.117$); and 5 year OS was 14% for standard risk patients and 8% for high-risk patients ($p = 1.000$) (Figure 2).

For those patients who received surgical resection, 1 year OS survival was 70% as compared to 40% for those who did not ($p = 0.069$); 2 year OS was 30% for surgical resection and 10% without surgical resection ($p = 0.582$) and 5 year OS was 20% for surgical resection and 0% without surgical resection ($p = 0.473$) (Figure 2).

Discussion

Hepatoblastoma is a malignancy that has been documented to have a poorer prognosis in LMICs where presentation is often late with advanced disease and resources to effectively treat the disease are limited.[17] In our study, the majority of the children presented with advanced stages of the disease with high-risk classification implying a delay in presentation and diagnosis.

The incidence of HB in this study was 1.2% (25 cases of HB out of 2076 admissions). This is lower than that observed in the literature. In a large cohort study published in Egypt in 2021, the incidence was noted to be 1.4% (208 children with HB out of 14,808 admissions with cancer).[17]

A higher median age of 24 months was noted in this study in contrast to other studies. Median age at presentation was 14 months in Turkey[8] and 12 months in the Nashville US, India and Cairo studies.[17,18,19] In a study conducted in Red Cross War Memorial Children's Hospital, in 2014, the median age was noted to be 12.8 months.[20] Age at presentation

TABLE 1: Clinical characteristics, investigation and results (N = 20).

Variable	Frequency	%
Age (years)		
0–1	5	25
> 1–2	8	40
> 2–3	5	25
> 3–4	2	10
> 4	0	0
Sex		
Male	13	65
Female	7	35
Nutritional status		
Under-nourished	7	35
Normal	12	60
Over-nourished	1	5
Gestational age (weeks)		
< 37	4	20
≥ 37	16	80
ICU admission in neonatal period		
Yes	3	15
No	17	85
Congenital abnormality or syndrome		
Yes	2	10
No	18	90
Family history of cancer		
Yes (liver cancer)	1	5
No	19	95
Symptoms at presentation		
Abdominal swelling	20	100
Abdominal pain	3	15
Loss of appetite	3	15
Weight loss	3	15
Jaundice	2	10
Investigations and results		
Alpha fetoprotein (AFP) (ng/mL)		
< 100	0	0
100–1 000 000	17	85
> 1 000 000	3	15
Haemoglobin (Hb) (g/dl)		
< 10	16	80
≥ 10	4	20
Platelets (cells/mcL)		
< 450	5	25
≥ 450	15	75
Alanine transferase (ALT) (U/L)		
< 40	13	65
≥ 40	7	35
Aspartate transferase (AST) (U/L)		
< 60	7	35
≥ 60	13	65
HIV testing		
Negative	19	95
Positive	0	0
Not documented	1	5
Radiological testing		
CXR	8	40
Abdominal sonar	10	50
CT scan	20	100
MRI	3	15
Biopsy		
Done	15	75
Not documented	5	25

Table 1 continues →

TABLE 1 (Continues...): Clinical characteristics, investigation and results (N = 20).

Variable	Frequency	%
Biopsy methods (n = 15)		
Surgical resection and/or open biopsy	4	27
Tru-cut	8	53
Fine needle	3	20
Histology (n = 15)		
Epithelial	11	73
Mixed epithelial and mesenchymal	4	27

ICU, intensive care unit; HIV, human immunodeficiency virus; CXR, chest X-ray; CT, computed tomography; MRI, magnetic resonance imaging.

TABLE 2: Staging and outcomes (N = 20).

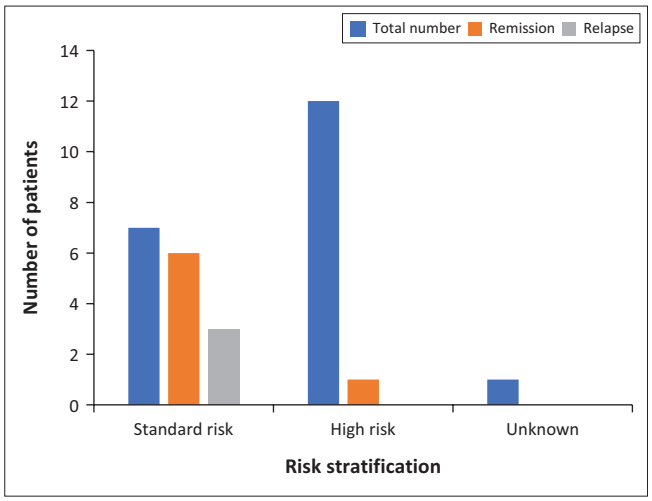
Variable	Frequency	%
Pretext staging		
I	2	10
II	3	15
III	3	15
IV	11	55
Not documented	1	5
Surgical staging		
I	7	35
IV	3	15
Not documented	10	50
Treatment at UAH		
Chemotherapy	19	95
Surgical resection	10	50
TACE	0	0
Liver transplant	0	0
Chemotherapy drugs		
PLADO	18	90
SUPER PLADO	1	15
Survival		
1 year overall survival	11	55
2 year overall survival	4	20
2 year event free survival	2	10
5 year overall survival	2	10
5 year event free survival	2	10

PLADO, Cis-Platinum + Doxorubicin combination; Super PLADO, Cis-platinum + Doxorubicin + Carboplatin combination; UAH, Universitas Academic Hospital; TACE, Trans Arterial-Chemo-embolisation; PRETEXT, PRE-Treatment EXTent.

younger than 3 years old has been linked with better prognosis in the literature[21,22] which was like our findings in this study. A larger and more robust study will be needed to confirm this finding.

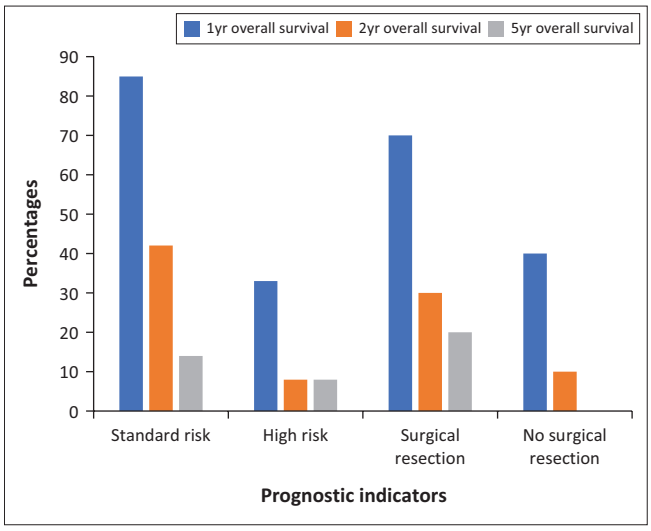
We also noted HB to predominate in male patients with a ratio of 1.9:1, which is consistent with the literature.[1,22,23] We, however, could not find evidence of higher mortality among male patients in the literature. In a study by Bassan J Allan et al in 2013, male patients were found to have higher survival rates.[24] More studies are needed to confirm this finding.

A minority of the children in our study, 20%, were born premature. This is not the case in high-income countries where prematurity and low birth weight have been found to increase the incidence of HB.[25,26] Of the four premature cases, one had a congenital brain malformation (absent corpus callosum) and presented with PRETEXT I and later developed acute lymphoblastic leukaemia (ALL); the other three children presented with advanced disease. Surveillance for intra-abdominal malignancies is important in premature babies.



Note: Showing that most standard risk patients had remission, while most high-risk patients had disease progression.

FIGURE 1: Relationship between risk and response to chemotherapy.



Note: Showing that overall survival was highest among standard risk patients and lowest among high-risk patients. Survival was also higher among patients who received surgical resection. Standard risk included PRETEXT I, II, III without annotation factors. High risk included tumour rupture, PRETEXT IV or any PRETEXT stage with annotation factors and distant metastasis.

PRETEXT, PRE-Treatment EXTent; yr, year.

FIGURE 2: Relationship between prognostic indicators and survival.

Two children in this study had congenital abnormalities: absent corpus callosum described above and the other was diagnosed with Simpson–Golabi–Behmel syndrome which is known in the literature to increase the risk of developing HB.[27] The child with Simpson–Golabi–Behmel syndrome was male and aged 2 years and 8 months at diagnosis. The AFP levels were high at 582,050 and he presented with advanced stage, that is, PRETEXT IV + lung metastasis + invasion of the portal vein and inferior vena cava. He did not get surgical resection but had a tru-cut biopsy which showed macrotrabecular histological subtype. He received PLADO for 6 X cycles and demised after 1 year of diagnosis. This highlights the need for surveillance of HB and other malignancies in these subsets of patients.

Undernutrition is still a burden in LMICs[16], and a third of our patients (n = 7) were noted to be under-nourished. Most of them (57%) presented with advanced stage of disease with PRETEXT IV and did not get surgical resection with 100% mortality.

We observed that all the children in our study presented with abdominal distension which is noted to be the most common clinical feature in the literature.[7] This underscores the need to take abdominal distension in young children very seriously especially in the clinics and primary health care facilities.

Although AFP levels less than 100 ng/mL have been identified as a poor prognostic feature in the literature[7], all the children in this study had an elevated AFP level of more than 100 ng/mL but OS was still lower than in the literature. A major confounding factor was likely the high-risk staging at presentation in the majority.

Most of the children had thrombocytosis (85%; n = 17) and presented with advanced stage of disease. In a study conducted by Archana et al in 2019 in India, thrombocytosis was also noted in the overwhelming majority, 90% of the patients.[28] Thrombocytosis has been associated with poor prognosis.[22] However, in our study, two out of the three patients with platelets of less than 450 cells/mcl presented with PRETEXT IV + lung metastasis and had poor outcomes as well. More studies are needed to establish an association between thrombocytosis and poorer outcomes.

Distant metastasis at the time of diagnosis has been described as a negative prognostic indicator.[29] In our study, lung metastasis was the most common annotation factor, which is consistent with the literature as the commonest site of metastasis.[30,31] In a study by Tian Zhi et al, lung metastasis accounted for 80.3% of distant metastasis[22] as compared to 100% in our study. One child also had brain metastasis and blood vessel invasion concurrently which signified late presentation and diagnosis when spread had already occurred.

In a study conducted in China in 2020, the most common PRETEXT stage at presentation was II (38.7%) and pretext IV was only 3.2%[29], while in our study, the most common PRETEXT stage was IV, 55% (n = 11), conferring high risk and negatively affecting outcomes. One child presented with spontaneous tumour rupture with accompanying haemorrhagic shock and demised within 5 days of presentation.

In the literature, the foetal histological subtype has been associated with the most favourable outcomes.[32] However, when incompletely resected, the prognostic effect was not demonstrable.[33] We had two long-term survivors in our study: (1) the one had foetal + embryonal subtype and (2) the other had mixed epithelial subtype. Out of the children who demised, the longest lived for 3 years and 7 months had foetal + mixed epithelial subtype. This correlates with the study by Fan Li et al which showed that recurrence free rates were higher in children with mixed pathological subtypes.[34] Studies of larger cohorts are needed to correlate survival with the histological subtypes.

TABLE 3: Relationship between prognostic indicators and mortality.

Prognostic factor	%	Mortality	Total number	Fischer's Exact Test (*p*-value)
Age (years)	-	-	-	0.530
< 3	73.3	11	15	-
≥ 3	100	5	5	-
Sex	-	-	-	0.101
Male	92.3	12	13	-
Female	54.1	4	7	-
Distant metastasis	-	-	-	0.591
Yes	88.9	8	9	-
No	72.7	8	11	-
PRETEXT staging	-	-	-	0.262
I, II, III	62.5	5	8	-
IV	90.9	10	11	-
Risk	-	-	-	0.117
Standard risk	57.14	4	7	-
High risk	91.67	11	12	-
Surgical resection	-	-	-	0.086
Yes	60	6	10	-
No	100	10	10	-

PRETEXT, PRE-Treatment EXTent.

Super PLADO, 5-fluorouracil and irinotecan were used for relapses and refractory HB which is a similar practice in other centres.[35] Serial echocardiography (ECHO) and renal function tests were conducted to monitor for toxicity but there was no record of hearing tests in our study. High-frequency irreversible hearing loss has been noted in the literature to be the most common side effect of cisplatin chemotherapy.[36] In a study conducted in France in 2021, ototoxicity was noted to be the most concerning long-term treatment-related morbidity. They recommended hearing tests to be performed at least once within 5 years after diagnosis.[36]

A study by Fan Li et al in 2021 showed that 71.1% of patients achieved initial complete remission, and they were standard risk patients (PRETEXT II and III) with a relapse rate of 30%.[34] Also, recurrence free survival rates were higher in children younger than 3 years and standard risk HB (PRETEXT II and III, no metastasis or vascular invasion).[34] In another study by T.Y Wang et al in China in 2008, 91% of the patients achieved remission.[37] In our study, only a third of patients, 35%, achieved initial complete remission which is lower than in the literature. Most of them (71%) had PRETEXT I staging, 5% PRETEXT II and 5% PRETEXT III, and only PLADO was used as chemotherapy. Three of these children (42%) relapsed, did not respond to second line and/or extended course of PLADO chemotherapy and subsequently demised; two abandoned treatments after 1 year and two were reportedly still alive at the time of the study (Table 3). Therefore, this study revealed that children with standard risk (PRETEXT stage I, II, III with no metastasis) were more likely to go into remission than high-risk patients. Our relapse rate is, however, higher than in the literature.

The lack of surgical resection of the tumour is a predictor of shorter OS.[4] Most patients in our study could not be offered surgical resection because they were too unstable due to advanced disease signifying late presentation and diagnosis. More so, 20% (n = 4) of them died within 2 months of the diagnosis. This greatly impacted survival because all 10 (50%) of them who did not have surgical removal of the tumour had died by the third year of diagnosis. This correlates with a study conducted in Malawi in 2014 which showed 100% mortality in children who had incomplete or no surgical removal of the tumour.[11] In a study conducted in India in 2020, children who had surgical resection showed 1 year OS of 100% and 5 year OS of 83%.[38] In our study, the children who had surgical resection had 1 year OS of 70%. Survival will therefore improve if patients present with less advanced stages of the disease, and the whole tumour is successfully resected.

In our study, no patients received LT even though liver transplantation has been associated with higher survival outcomes in the literature.[39] Liver transplantation has been described as the preferred treatment modality for HR–HB[38] of which most of our patients were. In a study conducted in Turkey in 2022, it was observed that 33% of patients, in whom surgical resection was not possible, underwent LT and they achieved 75% survival.[8] Also, in a study conducted in Seoul, South Korea in 2021, LT was observed to have a strong positive impact on the OS of patients with PRETEXT IV from 20% to 94%.[40] A study conducted at Red Cross War Memorial Hospital in 2004 showed that the development of liver transplant programme led to marked improvement in survival from 14% to 80%.[20,41] The 1 year OS for patients with PRETEXT IV in our study was noted to be 30%.

In South Africa, currently, there are only two liver transplant centres namely, Red Cross War Memorial Hospital and The Wits Donald Gordon Medical Centre. The latter has the largest dedicated paediatric liver transplant unit in sub-Saharan Africa. While LT in HB is a potentially curative option for some patients, it is not always the best choice. Factors such as chemotherapy response, tumour resectability, presence of metastases and the patient's overall health play a role in the decision to proceed with transplantation or not. The above-mentioned factors played a considerable role in why it was mostly not considered as a treatment option. It emphasises the need to seek treatment in the early stages of the disease and complete resection of the tumour as the best chance of longer survival.

The median survival time in months in this study was 10 months which was significantly lower than that documented in literature: 17.86 months in Egypt in 2021[17] and 45 months median follow-up time in Germany where children presented with less advanced disease.[4]

At the time of this study, 16 (80%) of the children had died even though HB has now been described as one of the curable malignancies in the paediatric population.[42] Our study revealed a 1 year OS of 70% and a 5 year OS of 10% which is quite low compared to the findings of a study conducted in China in 2021 which showed 1 year OS of 95.3% and a 5 year

OS of 79.8%.[22] Also, the study conducted in Egypt by Soliman et al published in 2021 showed a 1 year OS of 73.8% and a 5 year OS of 51.3%.[17]

Limitations

This study has the following limitations:

- Retrospective study which relied on accurate record keeping and some information could not be obtained.
- Single-centre study in which the findings may not be generalisable.
- Small sample size as HB is a rare disease. More robust studies with larger number of patients will be needed to better corelate prognostic indicators with survival.

Recommendations

The study puts forward the following recommendations:

- Need for more education and awareness especially in the primary healthcare level on the red flags of HB which commonly presents with abdominal swelling.
- Prompt referral of children with HB in less advanced stages of disease to oncology centres which will increase chances of complete resection of the tumour and improved survival.
- Liver transplant services, although scarce and indicated for other subsets of patients, will aid in improving outcomes in high-risk HB.
- Regular hearing testing to detect possible ototoxicity.
- Accurate record keeping and documentation of medical notes as some of the information could not be obtained.
- Need for more research in South Africa on incidence and outcomes of HB to compare data and target interventions aimed at improving clinical management and survival.

Conclusion

This study has demonstrated that advanced stage of the disease which is suggestive of delayed presentation and diagnosis is a huge setback in achieving good outcomes. Also, the lack of surgical intervention like liver resection is the major factor in lower survival rates. Liver transplantation is fraught with its own setbacks like unavailability and difficulties to access. Early presentation is critical, and close attention must be paid to children under 3 years presenting with abdominal distension caused by the abdominal mass and elevated AFP levels. Early imaging of the abdomen and urgent referral to an oncology centre might improve outcomes.

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Competing interests

The authors, Helen Okafor and Jan du Plessis, declare that they have no financial or personal relationships that may have inappropriately influenced them in writing this article.

CRediT authorship contribution

Helen Okafor: Conceptualisation, Data curation, Formal analysis, Investigation, Methodology, Writing – original draft, Writing – review & editing. Jan du Plessis: Conceptualisation, Funding acquisition, Supervision, Validation. All authors reviewed the article, contributed to the discussion of results, approved the final version for submission and publication and take responsibility for the integrity of its findings.

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Data availability

The data that support the findings of this study are not openly available and are available from the corresponding author, Helen Okafor, upon reasonable request.

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