Child's Nervous System
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ORIGINAL ARTICLE

Understanding the trajectory of research efforts in atypical teratoid rhabdoid tumors: a bibliometric analysis of the 50 most impactful studies to date

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Abstract

Purpose Atypical teratoid/rhabdoid tumor (ATRT) is a highly malignant embryonal tumor of the central nervous system (CNS) that occurs predominantly in children. More is being discovered about this disease to improve understanding and outcomes. The aim of this analysis was to evaluate citation and other bibliometric characteristics of the 50 most cited articles in the contemporary literature in order to better model the trajectory of our current efforts.

Methods Elsevier's Scopus database was searched for the 50 most cited articles about ATRT. To look for trends, earliest 25 articles were separated from the latest 25 articles and then were compared. Various bibliometric parameters were summarized and compared using Pearson's chi-square and Mann–Whitney *U* tests.

Results The 50 most cited articles were published between 1990 and 2016, from 5 unique countries in 29 unique journals, with genetic and retrospective observational cohort studies the most common design (n = 11 each). Overall median values were as follows: citation count, 145.4 citations (range, 67–626); citation rate per year, 11.7 (range, 3.5–51.4); number of authors 12 (range, 1–95); with 32 (64%) originating from the USA. Compared with older articles, newer articles had statistically lower citation counts (101.8 vs 189.0; $P < 0.01$), higher number of authors (17.3 vs 6.6; $P < 0.01$), and were less likely published from the USA (40% vs 88%; $P < 0.01$)

Conclusions The 50 most cited articles about ATRT were characterized in this analysis. There was a distinct focus in these studies on the genetic composition and consequences of these tumors. Trends over time suggest greater impact will be had in highly collaborative efforts worldwide. Moving forward, it will be of great interest to see how the findings of these basic science finding will translate into future clinical studies.

Keywords Pediatric · Atypical teratoid rhabdoid tumors · ATRT · Brain cancer · Bibliometric · Most cited

Introduction

Atypical teratoid/rhabdoid tumor (ATRT) is a highly malignant embryonal tumor of the central nervous system (CNS) that accounts for 16% of all pediatric embryonal CNS tumors [1]. The incidence of ATRT in children 0–4 years of age is 0.32 per 100,000 population, which is significantly higher than the incidence of 0.03 per 100,000 population seen in children 5–9 years of age[1, 2]. ATRT is most commonly seen in children 0–2 years of age, and the incidence markedly decreases after the age of 2 [3]. By the 2007 World Health Organization (WHO) classification of CNS tumors, ATRT was declared as one of three major CNS embryonal tumors, and it was further classified as a WHO grade IV malignancy which is where it has remained [3, 4]. This corresponds to the very poor prognosis of ATRT across all ages, with 6-month,

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1-year, and 5-year expected survival rates of 65%, 47%, and 28% respectively [3]. The prognosis for children < 3 years of age, however, is even more dismal. The overall 2-year survival rate for patients 1–3 years of age is about 37%, and for patients < 1 year of age, the 2-year survival rate is only about 22% [3].

Diagnosis of ATRT is predominantly based on morphology, light microscopy, and immunohistochemistry. Histologically, the tumor often presents as sheets or nests of rhabdoid cells. In addition to this clustering of rhabdoid cells, the tumor may also present with malignant mesenchymal and/or epithelial components, with or without neuroepithelial fields [5]. In addition to this histology, the 2016 WHO classification of CNS tumors further defined ATRT by alterations of the *hSNF5/INI-1* gene on chromosome 22, and this abnormality has proven to be diagnostically valuable [2].

Given the predilection of this tumor to affect children, treatment is incredibly challenging. Radiation therapy has shown to be effective against ATRT tumors [6]; however, it is understandably avoided in children < 3 years of age because of the associated deleterious, long-term neurocognitive sequalae [7]. Additionally, ATRT's rarity and high level of malignancy have made it extremely difficult to establish a standard curative chemotherapeutic regimen [8, 9]. Correspondingly, there is a need for more consistency in our current approach to managing ATRT to make meaningful advances in the field, both molecularly and clinically. To build this understanding, we depend upon academic literature and impactful research to share and receive information about this rare tumor. The objective of this analysis was to evaluate the bibliometrics of the 50 most cited ATRT articles in the current literature. Doing this allows us to examine our current understanding of this tumor, and with this understanding, the trajectory upon which we can continue to advance our efforts to improve treatment paradigms.

Methods

Search strategy

The search strategy was designed to capture all relevant published indexed articles referring to ATRT. An electronic search was performed using Scopus in April 2020. Elsevier's Scopus contains indexed articles from approximately 22,000 journals and captures one of the widest range of scientific articles and most robust citation data amongst all electronic databases [10]. The database was searched and screened for "ATRT" and "atypical teratoid rhabdoid tumor" in the Title, Abstract, and Keywords. Articles were sorted into descending citation count order, and the first 50 articles that satisfied all selection criteria were included. Selection criteria required articles to (1) describe a clinical or basic science aspect of ATRT, (2) with ATRT the main focus or majority of tumors described. Broad central nervous system tumor classifications were not considered specific enough for ATRT. Any discrepancies were resolved by discussion and consensus between authors. Publications were limited to the English language.

Data extraction

The following validated bibliometric parameters were extracted: article title, authors, journal, Scopus citations, year, citation count, number of authors, first and senior author names, and country and institution of corresponding author. In the case of studies originating from the United States (US), state was also recorded. A journal was considered surgical if its primary focus was surgery, otherwise it was considered clinical (i.e., nonsurgical). Surgical approaches (open and endovascular) were recorded when described by an article.

Statistical analyses

Comparative analyses were conducted using chi-square exact test and Wilcoxon rank-sum test for categorical and continuous data, respectively. Statistical significance was set at two-sided $p < 0.05$. All statistical analyses were performed using STATA 14.1 (StataCorp, College Station, Texas).

Results

Study characteristics

An initial search of Scopus yielded 1049 articles, and the 50 most cited articles were then identified successfully (Supplementary Table 1). The 10 most cited articles [11–20] are summarized in Table 1. All articles were published between 1990 and 2016, from 5 unique countries in 29 unique journals, with 12 articles published under the open access model. In terms of article design, genetic, and retrospective observational cohort studies were the most common ($n = 11$ each), followed by reviews ($n = 8$) (Fig. 1). In terms of article focus, there were 26 articles that focused on clinical, patient outcomes, and 24 articles that reported basic science outcomes.

Citations

Overall, mean citation count and citation rate were 145.4 citations (range, 67–626) and 11.7 citations/year (range, 3.5–51.4) respectively (Fig. 2). The most cited article by count was the genetic study by Biegel et al. [11] from the Children's Hospital of Philadelphia, USA, titled "Germ-line and acquired mutations of INI1 in atypical teratoid and

Table 1 Summary of the ten most cited articles about ATRT

Citations									
No.	n	Rate (n/yr)	Year	First author	Senior author	Title	Journal	Country	PubMed ID
1	626	29.8	1999	Biegel JA	Fogelgren B	Germ-line and acquired mutations of INI1 in atypical teratoid and rhabdoid tumors	Cancer Research	US	9892189
2	573	23.9	1996	Rorke LB	Biegel JA	Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: Definition of an entity	Journal of Neurosurgery	US	8683283
3	358	51.1	2013	Knutson SK	Keilhack H	Durable tumor regression in genetically altered malignant rhabdoid tumors by inhibition of methyltransferase EZH2	Proceedings of the National Academy of Sciences of the United States of America	US	23620515
4	334	15.2	1998	Burger PC	Perlman EJ	Atypical teratoid/rhabdoid tumor of the central nervous system: A highly malignant tumor of infancy and childhood frequently mistaken for medulloblastoma: A Pediatric Oncology Group study	American Journal of Surgical Pathology	US	9737241
5	278	17.4	2004	Hilden JM	Biegel JA	Central nervous system atypical teratoid/rhabdoid tumor: Results of therapy in children enrolled in a registry	Journal of Clinical Oncology	US	15254056
6	268	16.8	2004	Judkins AR	Biegel JA	Immunohistochemical Analysis of hSNF5/INI1 in Pediatric CNS Neoplasms	American Journal of Surgical Pathology	US	15105654
7	268	14.9	2002	Biegel JA	Rorke LB	Alterations of the hSNF5/INI1 gene in central nervous system atypical teratoid/rhabdoid tumors and renal and extrarenal rhabdoid tumors	Clinical Cancer Research	US	12429635
8	253	16.9	2005	Tekautz TM	Gajjar A	Atypical teratoid/rhabdoid tumors (ATRT): Improved survival in children 3 years of age and older with radiation therapy and high-dose alkylator-based chemotherapy	Journal of Clinical Oncology	US	15735125
9	233	21.2	2009	Chi SN	Kieran MW	Intensive multimodality Treatment for children with newly diagnosed CNS atypical teratoid rhabdoid tumor	Journal of Clinical Oncology	US	19064966
10	186	10.3	2002	Zhang ZK	Kalpana GV	Cell cycle arrest and repression of cyclin D1 transcription by INI1/hSNF5	Molecular and Cellular Biology	US	12138206

rhabdoid tumors" with 626 citations published in 1999 in *Cancer Research*. The most cited article by rate was the in vivo study by Knutson et al. [13] Epizyme, Inc., USA, titled "Durable tumor regression in genetically altered malignant rhabdoid tumors by inhibition of methyltransferase EZH2" with rate of 51.1 citations/year published in 2013 in *Proceedings of the National Academy of Sciences of the United States of America.*

Year of Publication

The median year of publication was 2006 (range, 1990–2016), which was also the year of most articles ($n = 6$) (Fig. 3). The earliest article was the case series by Biegel et al. [21] from the Children's Hospital of Philadelphia, USA, titled "Monosomy 22 in rhabdoid or atypical tumors of the brain" published in 1990 in *Journal of Neurosurgery.* The latest article was the

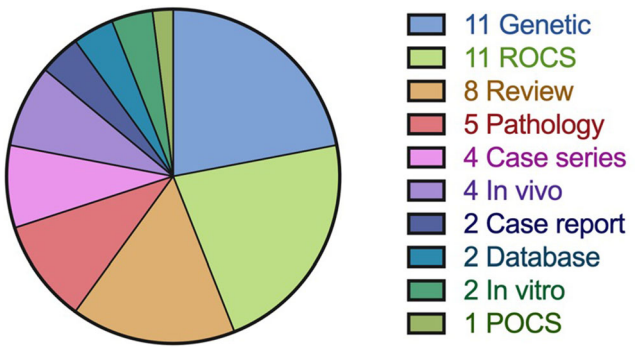

Fig. 1 Distribution of article content of articles. ROCS, retrospective observational cohort study; POCS, prospective observational cohort study

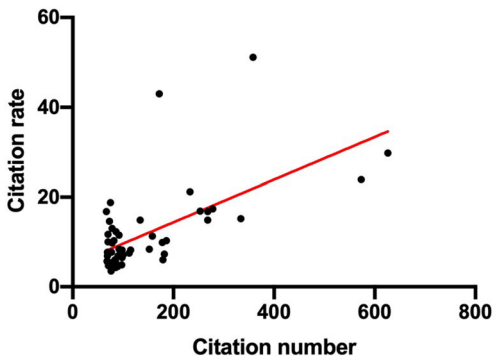

Fig. 2 Comparison of citation number and rate of articles with linear line of best fit

review by Fruhwald et al. [22] from the Children's Hospital Augsburg, Germany, titled "Atypical teratoid/rhabdoid tumors - Current concepts, advances in biology, and potential future therapies" published in 2016 in *Neuro-oncology*.

Authorship

There were 37 unique first authors and 35 unique senior authors. The mean number of listed authors across all articles was 12 (range, 1–95). The article with the greatest number of authors was the genetic study by Torchia et al. [23] from the University of Toronto, Canada, titled "Integrated (epi)-Genomic Analyses Identify Subgroup-Specific Therapeutic Targets in CNS Rhabdoid Tumors" published in 2016 in *Cancer Cell* with 95 coauthors. Jaclyn A. Biegel from the Children's Hospital of Philadelphia, United States, was the most prominent first and senior author across all included studies, with 7 first author and 8 senior author articles (Supplementary Table 1).

Countries of correspondence

Of the 5 unique countries that were listed as country of correspondence, the most contributions were made from the USA ($n = 32$) followed by Germany ($n = 9$), Canada ($n = 4$), Taiwan ($n = 3$) and Austria ($n = 2$). Within the USA, the most contributions were from PA ($n = 13$) followed by NY ($n = 4$), and TN ($n = 3$) (Supplementary Table 2).

Institutions

There were 26 unique institutions of correspondence (Supplementary Table 3). The most common institutions were the Children's Hospital of Philadelphia, USA ($n = 12$), University Hospital Münster, Germany ($n = 3$), Taipei Veterans General Hospital, Taiwan ($n = 3$), and St. Jude Children's Research Hospital, USA ($n = 3$).

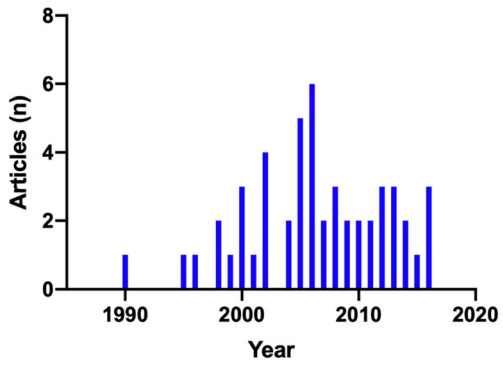

Fig. 3 Distribution of articles by year of publication

Journals

Articles were published in 29 unique journals (Supplementary Table 4). The most common journals were *American Journal of Surgical Pathology* ($n = 4$), and then *Clinical Cancer Research*, *Genes Chromosomes and Cancer*, *Journal of Clinical Oncology*, and *Pediatric Blood and Cancer* (each $n = 3$).

Newer versus older articles

To assess any shifts over time, the earliest 25 articles were compared with the latest 25 articles (Table 2). Notably, this separation occurred at publication year 2006. Compared with older articles, newer articles had statistically lower citation counts (101.8 vs 189.0; $P < 0.01$), higher number of authors (17.3 vs 6.6; $P < 0.01$), and were less likely published from the USA (40% vs 88%; $P < 0.01$)

Discussion

The intention of this bibliometric analysis was to identify and examine the 50 most impactful ATRT studies, based on citation count, in the current academic literature. Overall, out of the 50 articles, there were 26 articles (52%) that concentrated on clinical, patient outcomes, and there were 24 articles (48%) that focused on basic science outcomes. This illustrates the equal importance of basic science and clinical research in our contemporary approach to treating and researching ATRT.

The most cited article was published in 1999 by Biegel and colleagues [11]. This paper set the stage for the genetic characterization of ATRT by illuminating *INI-1* as the mutated tumor suppressor gene majorly responsible for the pathogenesis of this tumor. Biegel has contributed greatly to the molecular and genetic understanding of ATRT, and she is the senior author of 3 of the top 10 most cited articles [12, 15, 16]. Her findings are still heavily relied upon today for the diagnosis and identification of ATRT, and they have inspired the basic science research of this field.

As Biegel and colleagues discovered, ATRT is genetically characterized by a loss of the *hSNF5/INI-1* gene on chromosome 22. The INI1 protein is a part of the SW1/SNF chromatin remodeling complex, and it plays an important role in the maintenance of mitotic spindle and cell cycle control [11]. This mutation was the subject of numerous basic science papers within our cohort [17, 20, 24–26], and it is a major molecular marker that is used to distinguish ATRT from other similarly presenting CNS tumors [25, 26]. This extensive focus on the mutated tumor suppressor gene highlights the large impact of genetic research on the field of ATRT. While our understanding of the genetic profile of ATRT has become

Table 2 Comparison of articles published before (Older group) and after (Newer group) 2006

Parameter	Older group (n = 25)	Newer group (n = 25)	P-value
Quantitative (mean, range)			
Citations, number	189.0 (71-626)	101.8 (67-358)	**<0.01**
Citations, rate	10.2 (3.5-29.8)	13.4 (5.2-51.1)	0.29
Number of authors	6.6 (1-13)	17.3 (1-95)	**<0.01**
Qualitative (n, %)			
Article focus			0.57
Clinical outcomes	14, 56%	12, 48%	
Basic science outcomes	11, 44%	13, 52%	
Study design			
Retrospective observational cohort	6, 24%	5, 20%	0.73
Genetic	6, 24%	5, 20%	0.73
Published from United States	22, 88%	10, 40%	**<0.01**
Open access articles	4, 16%	8, 32%	0.19

more substantial, there has been limited translation of this knowledge into targeted clinical studies. In order for significant strides to be made in regard to treating this aggressive neoplasm, research must shift from simply understanding the genetics of ATRT to understanding how to use this information in a clinical setting.

In terms of treatment investigation, a number of articles included in this analysis explored intensive ATRT therapies across cohort studies, the most common study design [18, 19, 27]. Currently, there is no standard curative regimen for ATRT, but these articles demonstrate a future trajectory for ATRT treatment. One such therapy strategy, published by Tekautz and collegues in 2005, utilized an intensive multimodality treatment with radiation therapy and high-dose alkylator–based chemotherapy [18]. Another treatment approach, published by Gardner and collegues in 2008, included intensive induction chemotherapy followed by high dose chemotherapy with stem cell rescue [27]. This approach avoids the use of radiation, which may be incredibly beneficial for patients < 3 years of age. While some strides have been made in terms of treatment exploration, there is still a scarcity of randomized clinical trials. This is a shortfall that will be difficult to remedy because of the rare and aggressive nature of this tumor; however, it is a goal that the ATRT field must continue to strive towards. The exact intensity and combinations of modalities for optimal ATRT therapy has yet to be discovered, and we believe that the contribution of clinical articles such as the ones discussed need to, and will continue to, grow in the future.

Many of the articles in this cohort examined ATRT as a diagnosis on a broader scale. They discussed its definition as an entity, its characteristic clinical features, and its prevalence and incidence [5, 12, 14, 22, 28, 29]. This highlights the historic need to profile ATRT appropriately, and while understanding ATRT as a whole is important, the emphasis on these studies illustrates the limited nature of current ATRT research. Our analyses are not a reflection of completed trends in ATRT research; rather, they are an exploration of where the field is now and a foreshadowing of where the field will progress to next. In order to achieve greater clinical understanding of ATRT, research will need to move away from general profiling and genetic studies and shift towards more extensive translational and clinical investigations.

Finally, our analysis demonstrated that newer, more impactful studies had a higher number of authors than older articles and were also less likely to be published in the USA. The increase in coauthor numbers in newer articles indicates that ATRT research is becoming more and more collaborative with greater multidisciplinary interaction. This growth in collaboration presumably increases the necessary time spent for article preparation and dissemination, as well as accumulation of meaningfully sized cohorts for study. This may be why the median year of publication for our study cohort was 2006, with the last article being published in 2016 by Fruhwald and colleagues [22]. The latter point regarding global publication may be a reflection of the worldwide dissemination of the early, impactful studies conducted by Biegel and colleagues from the Children's Hospital of Philadelphia, PA, in the USA, the institution with the greatest contribution to our current ATRT understanding both in the USA and the world. Since then, active research efforts have reached institutions in Germany, Taiwan, Canada, and Austria. It is likely that future global efforts will be necessary to overcome the current issue of limited sample sizes, and the multidisciplinary and multinational directives derived from these worldwide endeavors will further advance the field of ATRT research.

There are limitations to this study in its current form. It must be noted that utilizing citation numbers as a proxy for article impact is imperfect. Citation numbers may be more representative of academic use rather than clinical or

translational impact. That being said, about half of our cohort represented clinical papers, which argues the point that citation count has predictive value for clinical usefulness. Furthermore, there are other bibliometric measures that can be assessed, such as journal impact factor, which can influence the article's exposure and accessibility to the general audience. For article sourcing with citation data, we utilized the Scopus database, which despite its more robust citation data, is more limited in terms of articles published prior to 1995 compared with other databases [10]. Encouragingly, our analysis identified an article [21] published in 1990 within the 50 most cited articles suggesting this limitation may not have impacted our results greatly, particularly given the more recent nature of ATRT research per our findings. Finally, it is important to remember that citation numbers are constantly growing, and it takes time for a newer article to accrue citations. This paper presents the 50 most cited articles for ATRT at this current period of time, but we predict this list will change over time as citation numbers evolve for more recent studies. Correspondingly, we encourage readers to interpret our trends as a snapshot in time about ATRT research and as with citation numbers, evolve over time.

Conclusions

In this bibliometric analysis, we identified the 50 most cited articles about ATRT and showed that ATRT research is still in a growing phase, with emphasis placed on understanding the genetic, clinical, and epidemiological facets of the tumor. Clinical articles regarding treatment of ATRT constituted a smaller subset of our article cohort, depicting the need for the development of effective therapy regimens. Hopefully, as our scientific understanding of ATRT grows, and as research efforts become more collaborative and global, we will see translations of that knowledge into future treatment paradigms.

Compliance with ethical standards

Conflict of interest The authors report no conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

1. Ostrom QT, Cioffi G, Gittleman H, Patil N, Waite K, Kruchko C, Barnholtz-Sloan JS (2019) CBTRUS Statistical report: primary brain and other central nervous system tumors diagnosed in the United States in 2012–2016. Neuro-Oncology 21:v1–v100
2. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, Ohgaki H, Wiestler OD, Kleihues P, Ellison DW (2016) The 2016 World Health Organization classification of tumors of the central nervous system: a summary. Acta Neuropathol 131:803–820
3. Ostrom QT, Chen Y, MdB P, Ondracek A, Farah P, Gittleman H, Wolinsky Y, Kruchko C, Cohen ML, Brat DJ, Barnholtz-Sloan JS (2014) The descriptive epidemiology of atypical teratoid/rhabdoid tumors in the United States, 2001-2010. Neuro-Oncology 16:1392–1399
4. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, Scheithauer BW, Kleihues P (2007) The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol 114:97–109
5. Packer RJ, Biegel JA, Blaney S, Finlay J, Geyer JR, Heideman R, Hilden J, Janss AJ, Kun L, Vezina G, Rorke LB, Smith M (2002) Atypical teratoid/rhabdoid tumor of the central nervous system: Report on workshop. J Pediatr Hematol Oncol 24:337–342
6. Chen YW, Wong TT, Ho DM, Huang PI, Chang KP, Shiau CY, Yen SH (2006) Impact of radiotherapy for pediatric CNS atypical teratoid/rhabdoid tumor (single institute experience). Int J Radiat Oncol Biol Phys 64:1038–1043
7. Anderson DM, Rennie KM, Ziegler RS, Neglia JP, Robison LR, Gurney JG (2001) Medical and neurocognitive late effects among survivors of childhood central nervous system tumors. Cancer 92:2709–2719
8. Ginn KF, Gajjar A (2012) Atypical teratoid rhabdoid tumor: current therapy and future directions. Front Oncol 2:114
9. Finkelstein-Shechter T, Gassas A, Mabbott D, Huang A, Bartels U, Tabori U, Janzen L, Hawkins C, Taylor M, Bouffet E (2010) Atypical teratoid or rhabdoid tumors: improved outcome with high-dose chemotherapy. J Pediatr Hematol Oncol 32:e182–e186
10. Falagas ME, Pitsouni EI, Malietzis GA, Pappas G (2008) Comparison of PubMed, Scopus, Web of Science, and Google Scholar: strengths and weaknesses. FASEB J 22:338–342
11. Biegel JA, Zhou JY, Rorke LB, Stenstrom C, Wainwright LM, Fogelgren B (1999) Germ-line and acquired mutations of INI1 in atypical teratoid and rhabdoid tumors. Cancer Res 59:74–79
12. Rorke LB, Packer RJ, Biegel JA (1996) Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: definition of an entity. J Neurosurg 85:56–65
13. Knutson SK, Warholic NM, Wigle TJ, Klaus CR, Allain CJ, Raimondi A, Scott MP, Chesworth R, Moyer MP, Copeland RA, Richon VM, Pollock RM, Kuntz KW, Keilhack H (2013) Durable tumor regression in genetically altered malignant rhabdoid tumors by inhibition of methyltransferase EZH2. Proc Natl Acad Sci U S A 110:7922–7927
14. Burger PC, Yu IT, Tihan T, Friedman HS, Strother DR, Kepner JL, Duffner PK, Kun LE, Perlman EJ (1998) Atypical teratoid/rhabdoid tumor of the central nervous system: a highly malignant tumor of infancy and childhood frequently mistaken for medulloblastoma: a Pediatric Oncology Group study. Am J Surg Pathol 22:1083–1092
15. Hilden JM, Meerbaum S, Burger P, Finlay J, Janss A, Scheithauer BW, Walter AW, Rorke LB, Biegel JA (2004) Central nervous system atypical teratoid/rhabdoid tumor: results of therapy in children enrolled in a registry. J Clin Oncol 22:2877–2884
16. Judkins AR, Mauger J, Ht A, Rorke LB, Biegel JA (2004) Immunohistochemical analysis of hSNF5/INI1 in pediatric CNS neoplasms. Am J Surg Pathol 28:644–650
17. Biegel JA, Tan L, Zhang F, Wainwright L, Russo P, Rorke LB (2002) Alterations of the hSNF5/INI1 gene in central nervous system atypical teratoid/rhabdoid tumors and renal and extrarenal rhabdoid tumors. Clin Cancer Res 8:3461–3467
18. Tekautz TM, Fuller CE, Blaney S, Fouladi M, Broniscer A, Merchant TE, Krasin M, Dalton J, Hale G, Kun LE, Wallace D, Gilbertson RJ, Gajjar A (2005) Atypical teratoid/rhabdoid tumors (ATRT): Improved survival in children 3 years of age and older with radiation therapy and high-dose alkylator-based chemotherapy. J Clin Oncol 23:1491–1499

19. Chi SN, Zimmerman MA, Yao X, Cohen KJ, Burger P, Biegel JA, Rorke-Adams LB, Fisher MJ, Janss A, Mazewski C, Goldman S, Manley PE, Bowers DC, Bendel A, Rubin J, Turner CD, Marcus KJ, Goumnerova L, Ullrich NJ, Kieran MW (2009) Intensive multimodality Treatment for children with newly diagnosed CNS atypical teratoid rhabdoid tumor. J Clin Oncol 27:385–389
20. Zhang ZK, Davies KP, Allen J, Zhu L, Pestell RG, Zagzag D, Kalpana GV (2002) Cell cycle arrest and repression of cyclin D1 transcription by INI1/hSNF5. Mol Cell Biol 22:5975–5988
21. Biegel JA, Rorke LB, Packer RJ, Emanuel BS (1990) Monosomy 22 in rhabdoid or atypical tumors of the brain. J Neurosurg 73:710–714
22. Frühwald MC, Biegel JA, Bourdeaut F, Roberts CWM, Chi SN (2016) Atypical teratoid/rhabdoid tumors - Current concepts, advances in biology, and potential future therapies. Neuro-Oncology 18:764–778
23. Torchia J, Golbourn B, Feng S, Ho KC, Sin-Chan P, Vasiljevic A, Norman JD, Guilhamon P, Garzia L, Agamez NR, Lu M, Chan TS, Picard D, de Antonellis P, Khuong-Quang DA, Planello AC, Zeller C, Barsyte-Lovejoy D, Lafay-Cousin L, Letourneau L, Bourgey M, Yu M, Gendoo DMA, Dzamba M, Barszczyk M, Medina T, Riemenschneider AN, Morrissy AS, Ra YS, Ramaswamy V, Remke M, Dunham CP, Yip S, Ng HK, Lu JQ, Mehta V, Albrecht S, Pimentel J, Chan JA, Somers GR, Faria CC, Roque L, Fouladi M, Hoffman LM, Moore AS, Wang Y, Choi SA, Hansford JR, Catchpoole D, Birks DK, Foreman NK, Strother D, Klekner A, Bognár L, Garami M, Hauser P, Hortobágyi T, Wilson B, Hukin J, Carret AS, Van Meter TE, Hwang EI, Gajjar A, Chiou SH, Nakamura H, Toledano H, Fried I, Fults D, Wataya T, Fryer C, Eisenstat DD, Scheinemann K, Fleming AJ, Johnston DL, Michaud J, Zelcer S, Hammond R, Afzal S, Ramsay DA, Sirachainan N, Hongeng S, Larbcharoensub N, Grundy RG, Lulla RR, Fangusaro JR, Druker H, Bartels U, Grant R, Malkin D, McGlade CJ, Nicolaides T, Tihan T, Phillips J, Majewski J, Montpetit A, Bourque G, Bader GD, Reddy AT, Gillespie GY, Warmuth-Metz M, Rutkowski S, Tabori U, Lupien M, Brudno M, Schüller U, Pietsch T, Judkins AR, Hawkins CE, Bouffet E, Kim SK, Dirks PB, Taylor MD, Erdreich-Epstein A, Arrowsmith CH, De Carvalho DD, Rutka JT, Jabado N, Huang A (2016) Integrated (epi)-genomic analyses identify subgroup-specific therapeutic targets in CNS rhabdoid tumors. Cancer Cell 30:891–908
24. Biegel JA, Kalpana G, Knudsen ES, Packer RJ, Roberts CW, Thiele CJ, Weissman B, Smith M (2002) The role of INI1 and the SWI/SNF complex in the development of rhabdoid tumors: meeting summary from the workshop on childhood atypical teratoid/rhabdoid tumors. Cancer Res 62:323–328
25. Judkins AR, Burger PC, Hamilton RL, Kleinschmidt-DeMasters B, Perry A, Pomeroy SL, Rosenblum MK, Yachnis AT, Zhou H, Rorke LB, Biegel JA (2005) INI1 protein expression distinguishes atypical teratoid/rhabdoid tumor from choroid plexus carcinoma. J Neuropathol Exp Neurol 64:391–397
26. Judkins AR (2007) Immunohistochemistry of INI1 expression: a new tool for old challenges in CNS and soft tissue pathology. Adv Anat Pathol 14:335–339
27. Gardner SL, Asgharzadeh S, Green A, Horn B, McCowage G, Finlay J (2008) Intensive induction chemotherapy followed by high dose chemotherapy with autologous hematopoietic progenitor cell rescue in young children newly diagnosed with central nervous system atypical teratoid rhabdoid tumors. Pediatr Blood Cancer 51:235–240
28. Lafay-Cousin L, Hawkins C, Carret AS, Johnston D, Zelcer S, Wilson B, Jabado N, Scheinemann K, Eisenstat D, Fryer C, Fleming A, Mpofu C, Larouche V, Strother D, Bouffet E, Huang A (2012) Central nervous system atypical teratoid rhabdoid tumours: the Canadian Paediatric Brain Tumour Consortium experience. Eur J Cancer 48:353–359
29. Athale UH, Duckworth J, Odame I, Barr R (2009) Childhood atypical teratoid rhabdoid tumor of the central nervous system: a meta-analysis of observational studies. J Pediatr Hematol Oncol 31:651–663