

Positional Plagiocephaly

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Abstract

In 1992, the American Academy of Pediatrics initiated a campaign “Back to Sleep” which implied to place the healthy neonates and infants in supine position to reduce Sudden Infant Death Syndrome. Due to this, the incidence of Sudden Infant Death Syndrome reduced. However, the prevalence of positional plagiocephaly drastically increased. Positional plagiocephaly is one of the leading cause of skull asymmetry. Positional Plagiocephaly is seen due to continuous pressure/external force on one part of the skull either left or right occiput and also referred to as deformational, non-synostosis, and occiput plagiocephaly. In addition to resulting in cosmetic concerns, it is also associated with adverse neurodevelopmental outcome such as motor, cognitive and language delays in infancy and toddlers. Later learning problems are manifested in school age. Early diagnosis and prompt management is the key in preventing the condition. This review article may help the health care professional to diagnose early and provide appropriate intervention services.

Keywords: *Positional Plagiocephaly (PP), Deformational Plagiocephaly, helmet therapy, non cranio-synostosis.*

1. Background

Positional plagiocephaly (PP) also known as deformational plagiocephaly or non - synostosis plagiocephaly one of the leading cause of skull asymmetry or abnormal shape of the head in infants (Rogers). While positional plagiocephaly has been known for many decades, major attention has been drawn to this condition over the past 2 decades (Clarren, 1981). Besides cosmetic effect pp may be

associated with motor, cognitive, language deficits in infancy, toddlers and learning problems in school age children (Bialocerkowski, Vladusic, & Howell, 2005). Understanding of etio-pathogenesis & treatment strategies is key to primary prevention of this condition. Also early recognition and prompt management make a significant difference to ultimate outcome (B. Collett, Breiger, King, Cunningham, & Speltz, 2005). This present article is meant for an overview of positional plagiocephaly for health care providers such as neonatologists, pediatricians, therapist and nurses who may see this condition and diagnose early, may accordingly advise and educate parents in preventing pp.

1.1 Definition, Types and Prevalence:

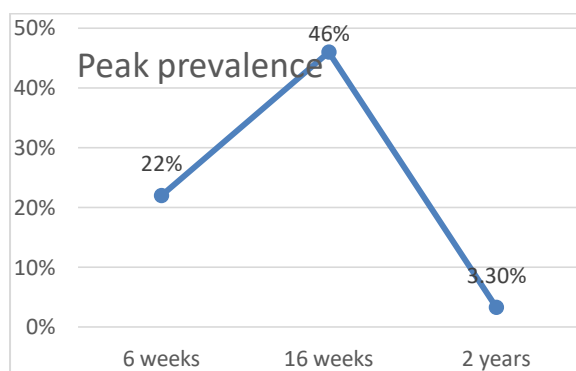
“Plagio” word derived from greek means “oblique” and cephalic means “head”. “Plagiocephaly” refers to an asymmetrical, flattened deformation of skull (Looman & Flannery). There are two types of plagiocephaly synostosis and non-synostosis plagiocephaly. *Non-synostosis* also known as positional plagiocephaly, deformational plagiocephaly, and occipital plagiocephaly (Looman & Flannery), (Shweikeh, Nuno, Danielpour, Krieger, & Drazin) is one sided occipital flattening either right or left. *Synostosis* also known as unilateral lambdoid suture is much less common, has genetic predisposition occurs due to closure of unilateral lambdoid suture (Huang, Mouradian, Cohen, & Gruss, 1998).

Incidence of pp has been increased after the campaign “Back to sleep” made by AAP force task in 1992. The campaign implied to place the healthy infants in supine position to decrease the sudden Infant Death Syndrome (SIDS) (J Kattwinkel, 1992). Incidence of SIDS was associated with a multifold increase in incidence of pp during the same period.

This was estimated to be equivalent to an increase of 21.2% from 1999 to 2007 (Kane, Mitchell, Craven, & Marsh, 1996).

Research reported on incidence of pp before and after “Back to sleep” campaign is shown in Table –I.

While pp may be noted at birth itself, the prevalence of pp is noticed to increase starting from 6 weeks – 7 weeks(16-22.1%) with a peak around 7 weeks to 4 months(46%) and then it decreases and stabilizes around 3.3% by about 2 years (Hutchison, Hutchison, Thompson, & Mitchell, 2004) fig -1.



2. Etio-Pathogenesis

Positional plagiocephaly can occur due to continuous external forces on infant’s growing skull either occurring prenatally or in early period of infancy.

2.1 Influence of intra uterine environment:

The risk of flattened skull is high in twins as much as in 56% than singletons because of the space constraint in the uterus and later the child preferentially continues to sleep in the flattened side resulting in the asymmetry of skull (Littlefield, Kelly, Pomatto, & Beals, 2002).

2.2 Right is more common than left:

Occiput of the fetus tends usually comes in contact against pelvis of mother in the last trimester for longer period. This results in almost continuous external pressure acting on the infants growing skull (Losee, Mason, Dudas, Hua, & Mooney, 2007),(Moss, 1997). After the birth, when placed for > 20hrs in supine position, the infant assumes a preferential position on right. Hutchison et al, found that spending <5 minutes per day in “tummy time” was significant risk factor for PP.

2.3 Cause and effect relationship of developmental problem and plagiocephaly:

The main question is whether developmental delay causes pp or is it a consequence of a skull deformity? The infants having prenatally or early postnatal developmental problem tends to demonstrate less motor activity(McKinney, Cunningham, Holt, Leroux, & Starr, 2009). In many other infants with PP, has different type of influencing factors such as supine sleeping, lack of prone timing (tummy time), low activity levels. PP does not cause developmental delay per se but it is a marker of elevated risk of developmental delays (B. Collett et al., 2005) fig -2

2.4 Prolonged stay in the intensive care unit:

Preterm babies and babies with developmental delays probably associated with prolonged stay in intensive care unit predisposes to PP (B. Collett et al., 2005).

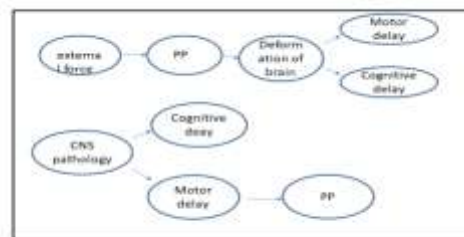


Figure -2

2.5 Risk Factors:

Recent studies identified risk factors responsible for positional plagiocephaly(Hutchison et al., 2004), (McKinney et al., 2009),(Bialocerkowski, Vladusic, & Wei Ng, 2008),(Ditthakasem & Kolar).The risk factors are shown in Table –II.

3. Management:

3.1 Presentation and diagnosis:

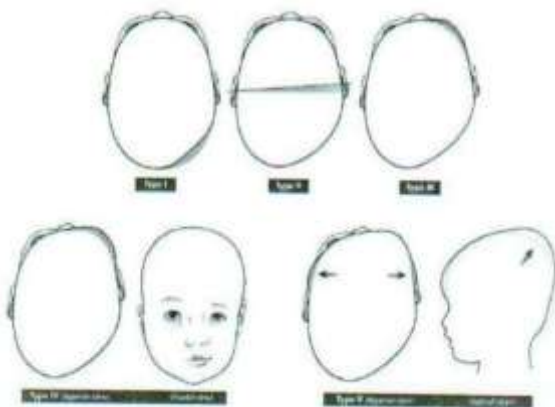
High index of suspicious, especially among at risk infants, along with prompt and early diagnosis is critical in achieving the good outcome in PP.A diagnosis requiring on a comprehensive history, careful physical examination along with specific tools including anthropometric calipers or digital devices with software analysis . Occasionally 3-D

imaging may be necessary (Flannery, Looman, & Kemper).

3.2 Clinical evaluation:

Every infant must be examined at birth. The infant with PP presents with unilateral flattening of occiput. Infants with skull asymmetry, a detailed examination from head to toe focusing on face and neck, the infant must be examined from anterior, posterior, lateral side and vertex must be performed. (23). Besides flattening of occiput, PP is accompanied with displacement of ipsilateral ear, contralateral bossing of occiput, ipsilateral frontal bossing and contralateral frontal flattening manifesting as a parallelogram shape. *Argenta*, provided a simple clinical classification of PP by four observational positions from anterior, posterior, lateral and from vertex. He classified them into five types (Argenta, David, & Thompson, 2004). Clinical classification is represented in the Fig 3.

Figure -3



Clinical Finding	Observation position	Type 1	Type 2	Type 3	Type 4	Type 5
Posterior Asymmetry	Directly posterior	Present	Present	Present	Present	Present
Ear Malposition	Directly above, seated in lap, fingers in ears	Absent	Present	Present	Present	Present
Frontal Asymmetry	Directly anterior	Absent	Absent	Present	Present	Present
Facial Asymmetry	Directly anterior	Absent	Absent	Absent	Present	Present
Temporal Bossing or Posterior Vertical Cranial Growth	Directly lateral	Absent	Absent	Absent	Absent	Present

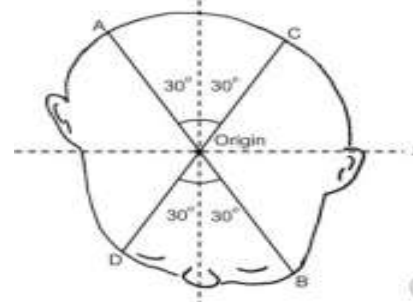
3.3. Specific evaluation for diagnosis:

Anthropometric calipers: The diagonal, width, length and circumference of the skull may be captured by applying standard cephalometric measurements using a tape measure. These anthropometric measurements taken on defined landmarks are

reliable but may be measured errors when baby is restless. Moss and Mortenson et.al. define the cranial vault asymmetry (CVA) as the difference between the largest and smallest diagonal diameter of the skull $AB-CD/AB$ where $AB > CD$ (fig—3). A CVA $< 3mm$ is regarded as physiological, $> 3mm$ and $< 12mm$ is regarded as mild to moderate symmetry and $> 12mm$ is regarded as moderate to severe asymmetry (Moss, 1997).

$CVAI = AB-CD/ AB \times 100$. Where AB is longer than CD . Fig- 4. Values below 3.5% are regarded as physiological. CVAI gives the objective assessment. The measurements may be helpful to plan for helmet therapy in severe cases where it is not responding to repositioning therapy (Kim, Park, Yang, & Yim)

Figure-4



Imaging: CT scanning/ imaging is done in some of the complex cases and helps to differentiate the synostosis and non-synostosis plagiocephaly. This helps to plan the surgical management /conservative management required to the index infant.

3.4 Treatment:

The age at which it is diagnose and the severity of the condition helps to decide strategy for treatment. The strategies for conservative treatment of PP includes repositioning of infants, mechanical adjustment exercises in mild cases and helmet therapy (cranial orthosis device) in moderate and severe condition (Persing, James, Swanson, & Kattwinkel, 2003). Repositioning of infant frequently avoids the continuous pressure on the skull and prevents flattening of the occiput (Flannery et al.). Repositioning can be given as one of the treatment in infants before four months of age (Persing et al., 2003) and improvement with repositioning is better before four months than the infants diagnosed later (Hutchison et al., 2004). If the infant with PP is associated with torticollis neck movement exercises may be beneficial to decrease the torticollis and also to eliminate this contributing factor for plagiocephaly.

Helmet therapy or cranial orthotic device seems to be beneficial in moderate to severe cases where repositioning fails after 6 months of age (Graham et al., 2005; Moss, 1997). However the challenges faced are compliance and frequent changing of helmet requiring to change every two to six months (Gump, Mutchnick, & Moriarty). The other challenge is that it is very costly, scalp infection, wearing for long hours. However, limited available evidence, does not recommend helmet therapy as a standard treatment and that has no helpful over repositioning (Goh, Bauer, Durham, & Stotland), (van Wijk et al.).

4. Neurodevelopmental outcome:

Though it is considered benign condition research studies reported, it may be associated with developmental problems represented in table- 3.

The above studies emphasize that PP does not cause developmental delays but developmental delays precede PP. ultimately, PP may serve as an efficient biological marker of elevated developmental risk (B. R. Collett et al.).

5. Prognosis:

Diagnosis and early treatment of positional plagiocephaly correlate positively with the outcome. Positional plagiocephaly can be fully resolved by means of conservative treatment (non -surgical) if recognized and treated early within 7/8 weeks from birth and can undergo significant improvement if diagnosed within first one year of age.

6. Prevention:

Educating the parents during the new born period (2-4 weeks) is the key for the prevention of the plagiocephaly. Repositioning of infant frequently is necessary to prevent the flattening of the occiput. Educating the parents about the importance of prone position and keeping upright under supervision when the baby is awake every day for three to four times for five to ten minutes or > 30 minutes per day (AAP 2011). This is known as "Tummy time" (John Person, 2003). This also helps the baby to achieve the early milestones in addition to the prevention of PP. The infant may be made to sleep or sit less in the car seat where occiput is on constant pressure (TR Littlefield, 2003). Health care providers plays a major role in educating the parents in teaching the positioning of infants for sleep, play and development when discharged.

7. Summary:

Positional plagiocephaly in an infant is a leading cause of skull shape deformation. PP has been increasing, reported as per after "Back to sleep" campaign launched by AAP 1992 strategy to decrease SIDS. This opportunity to understand the natural history and clinical outcome of PP, perinatal risk factors predisposing to PP. Positional plagiocephaly predisposes to adverse neurodevelopmental outcome. PP is causally related to adverse neurodevelopmental outcome. Primary prevention is vital to improve neuro-developmental outcome in high risk babies. Outcomes also depends on early diagnosing and prompt management. Principles of management are mainly repositioning, physical therapy exercises. Helmet therapy and surgery have no proven role in changing neurological outcome.

8. Conclusion:

Awareness of this condition which is not so benign needs to be created among all the health care providers. Educating the parents through clear guidelines regarding the recommendations of prone position for >30 min per day while awake, repositioning of the infant frequently and close monitoring is the key in preventing and managing this common condition. Simple clinical screening and examination helps to diagnose, to start early treatment to optimize the outcome. As a routine, health care providers such as pediatricians /nurses/ anganwadi workers should closely observe for infants with plagiocephaly during every health visit/immunization and prompt referral to early intervention services may be beneficial for the infants as point prevalence of PP is during 7 weeks to 12 weeks.

Table 1: Incidence of PP before and after back to sleep campaign

Author/year of publication	Study design & site	Before back to sleep campaign	After back to sleep campaign	% increase/fold
Kane AA/1996 (Kane et al., 1996)	chart review in a single tertiary craniofacial center			Sixfold
Branch, L.G./2015 (Branch et al.)	retrospective review by plastic surgeons at a tertiary medical center	9-178 patients between 1996-1999	316-576 patients between 2004-2007	390%
Sheu, SU/2011 (Sheu, Ethen, Scheuerle, & Langlois)	Descriptive epidemiologic study of time trends and a nested case-control study in the Texas Birth Defects Registry.	3 cases per 10,000 live births	28.8 cases per 10,000 live births	9 fold
1. Boere-Boonekam/2001 (Boere-Boonekamp & van der Linden-Kuiper, 2001) 2. Mawji, A/2013 (Mawji, Vollman, Hatfield, McNeil, & Sauve)	1. Prospective observational study at a tertiary care health centre 2. Prospective observational study in well-child clinic	8.2%	46.6%	38 %
1. Dunn PM /1974 (Dunn, 1974) 2. Turk AE/1996 (Turk, McCarthy, Thorne, & Wisoff, 1996)	1. Prospective observational study at a tertiary care health centre 2. Prospective observational study at craniofacial anomalies center	1 in 300	1 in 60	Five fold

Table 2: Etiological Factors

Socioeconomic factors	Obstetrics and perinatal factors	Infant risk factors	Infant care factors
Parental education		Male child	Continuously sleeping in supine position > 20hrs
Mothers age at birth	Breech presentation	first born child	Bottle feeding in one position
	Small for gestational age	Twin/triple	Infrequent tummy time > 30 min /day
	LBW	Positional preference of head	Placing in the car seat for longer period
	primiparous	Hydrocephalus	
	Congenital anomalies	Developmental delay	
		Slow achievements of motor milestones	
		Low activity levels	
		Limited head rotation	
		torticollis	

Table 3: Positional plagiocephaly associated with neurodevelopmental outcome

Author/year of publication	Study design	population	Test administered	Neuro-development outcome
Kordastanirouzheh et al.(Kordestani, Bard, Gurwitch, & Panchal, 2006)	Prospective study	110 infants with PP And 110 age matched controls	Bayleys scale	<i>Mental index</i> - 90%- normal 7% mild 3% severe delay <i>Psychomotor index</i> - 74% normal 19% mild delay 7% severe delay
Brent R collet et al 2011(B. R. Collett et al.)	Longitudinal study	227 infants with PP and 232 without pp	Bayley's scale at 7 months and 18 months	Scores were lower in Infants and toddlers with DP than without DP.
Miller and clarrein(Miller & Clarren, 2000)	Retrospective study	254 infants with pp at craniofacial center& Their sibilings as controls	Telephonicalinterview	39.7%- required help in school and other therapy --PP 7.7% - require help in controls
Eileen kennedy(Kennedy, Majnemer, Farmer, Barr, & Platt, 2009)	Prospective study	27 infants with pp & without pp (3-8 months of age)	Alberta infants motor performance(AIMS)	Scores-31.1 in infants with PP & 42.7 in without pp
BL Hutchison (Hutchison et al., 2004)	prospective	287 infants with pp(consequently)	Ages and stages questionnaire (ASQ)	One or more delays in 36% of infants with PP

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